DISSERTATION ON

RADIOLOGICAL EVALUATION OF SPINAL DYSRAPHISM USING CT AND MRI

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CERTIFICATE

This is to certify this dissertation titled **"RADIOLOGICAL EVALUATION OF SPINAL DYSRAPHISM USING CT AND MRI"** *submitted by Dr. S. KANAKA RAMESWARA KUMARAN, appearing for M.D. Branch VIII- RADIODIAGNOSIS degree examination in March 2007 is a bonafide record of work done by him under my direct guidance and supervision in partial fulfillment of regulations of the* Tamilnadu Dr.M.G.R.Medical University, Chennai. *I forward this to the Tamilnadu Dr. M.G.R Medical University Chennai, Tamil Nadu, India.*

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DECLARATION

I solemnly declare that this dissertation titled **"RADIOLOGICAL EVALUATION OF SPINAL DYSRAPHISM USING CT AND MRI"** has been done by me under the guidance and supervision of Prof.P.KUPPUSWAMY M.D,D.M.R.D, BARNARD INSTITUTE OF RADIOLOGY, MADRAS MEDICAL COLLEGE, CHENNAI. It is submitted towards partial fulfillment of requirements for the award of M.D.DEGREE (BRANCH VIII) RADIODIAGNOSIS to be held in March 2007 under the TAMIL NADU Dr.M.G.R.MEDICAL UNIVERSITY, CHENNAI, INDIA. This has not been submitted previously by me for award of any Diploma or Degree in any other University.

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CONTENTS

INTRODUCTION

Spinal dysraphism is the most common Neural tube defect in developing countries like India. The incidence varies from 0.5 to 11 per 1000 live births in different parts of our country, largely affecting the lower socioeconomic strata of the population.

The etiology of Neural tube defects is Multifactorial. The interaction of diverse factors related to Genetics, Nutrition and Environment play an important role in the etiopathogenesis. Some of the environmental factors that may contribute to Open Neural Tube Defects are uncontrolled maternal diabetes, and certain prescription medications. It has been proved that deficiency of essential vitamins especially Folic acid during pregnancy results in higher incidence of Neural tube defects which led to prophylactic supplementation of folic acid in the antenatal period.

Spinal dysraphism is a broad term encompassing a heterogeneous group of congenital spinal anomalies, which results from defective closure of the neural tube during early fetal life. Spinal dysraphism can be classified as Spina Bifida Aperta and Spina Bifida Occulta.

Spina bifida aperta is most common type of spinal dysraphism representing a serious congenital anomaly with severe Neurologic, Musculoskeletal, Genitourinary, and Bowel anomalies. It encompasses three forms namely Myelomeningocele, Myelocele and rarely Meningocele. Females show a higher incidence than Males and most of them present at birth and are immediately taken for surgical repair and hence are rarely imaged in unoperated cases.

Spina bifida occulta is characterized by minor Neurological manifestations and presents at a later age. Most distinct clinical findings are cutaneous stigmata like

Dermal dimple, Hemangioma, Cutis aplasia, Dermal sinus, or Hairy patch, Rudimentary tail (caudal appendage).

Segmentation anomalies of spine are a common feature of spinal dysraphism and along with muscle imbalances due to motor deficits result in Spinal curvature anomalies like Scoliosis, Kyphosis and Lordosis. Scoliosis is the most common type of spinal curvature anomaly.

Associated anomalies include Chiari malformations, Hydromyelia and Hydrocephalus.

Imaging plays a pivotal role in the diagnosis and management of spinal dysraphism.

The various imaging modalities available are

- **PLAIN RADIOGRAPH**
- **ULTRASONOGRAM**
- **CT**
- **MRI**

PLAIN RADIOGRAPH

Plain radiograph is the base line Investigation for diagnosing various vertebral anomalies like Spina bifida, Segmentation defects,spinal curvature anomalies, bony septum of Diastematomyelia , Spinal canal widening and Lumbosacral soft tissue swelling. How ever further characterization of the lesions require CT and MRI Imaging.

ULTRASONOGRAM

Spinal USG is a useful imaging modality for diagnosis of spinal dysraphism in infancy before maturation of the vertebral column.

Neurosonogram of the brain plays an important role in the diagnosis of associated anomalies of spinal dysraphism like hydrocephalus and chiari malformations.

COMPUTED TOMOGRAPHY

Computed tomography with Multiplanar reformation is an excellent modality for identification and characterization of vertebral segmentation defects, spinal curvature anomalies and bony septum in Diastematomyelia. Various forms of spinal dysraphism are diagnosed by CT based on the attenuation characteristics of their contents. Thus Meningocele shows fluid atteunation, Myelomeningoceles shows fluid and soft tissue attenuation and spinal lipomas show fat attenuation.CT is superceded by MRI in the detection of soft tissue and associated anomalies of spinal dysraphism like Hydromyelia, Hydrocephalus and Chiari malformations.

MAGNETIC RESONANCE IMAGING

MRI is the imaging modality of choice for characterizing the soft tissue spinal anomalies of Spinal dysraphism especially spinal cord. Meningomyelocele, Myelocele and Meningocele are evaluated according to the signal characteristics of their contents. Spinal Lipomas are best characterized using Fat Suppression Sequences. Both CT and MRI evaluates dorsal dermal sinus effectively. However MRI supercedes CT in further characterization of the tract. MRI best depicts fibrous septum in Diastematomyelia while bony septum is best demonstrated by CT. Further characterization into Split cord malformation I and II, location, extent of the hemicords, site of rejoining and associated anomalies are best demonstrated by MRI.

MRI best characterizes tethering of cord, spinal curvature anomalies, Chiari malformations, Hydromelia and Hydrocephalus.

Thus CT and MRI are complementary to each other in diagnosis and characterization of spinal dysraphism. Imaging also plays an important role in the postoperative follow-ups.

AIM OF THE STUDY

To assess the role of Helical CT and MRI in

- The identification of various forms of Spinal dysraphism.
- Characterization of the lesions and associated anomalies.
- Giving a composite diagnosis based on specific Imaging findings.

NORMAL ANATOMY

LUMBOSACRAL SPINE

LS spine is made up of

- ***Bony components***

Anterior elements and Posterior elements

- ***Ligaments***

  Anterior Longitudinal Ligament, Posterior Longitudinal Ligament, Ligamentum flavum etc

- ***Soft tissue components***

Epidural fat and veins

- ***Neural tissues and Meninges***

Spinal Cord, Conus Medullaris, Nerve Roots and meninges.

BONY COMPONENTS

ANTERIOR ELEMENTS

Vertebral bodies

There are 5 lumbar and 5 sacral segments with square shape. Superior and inferior endplates are covered by hyaline cartilage. The peripheral cortical bone is dense while the Inner medullary is trabeculated and contains marrow tissue.

Age	*Marrow type*	*T_1W*	*T_2W*	*T_1 Contrast*
<7yrs	Red marrow	Isointense	Isointense	Enhances
>7yrs	Yellow marrow	Hyperintense	Hypointense	No enhancement

Intervertebral discs

Intervertebral discs are bean shaped with a concave posterior margin except in S1. Nucleus pulposus shows high signal on T2 while annulus fibrosis shows low signal on T1 and T2 weighted sequences.

POSTERIOR ELEMENTS

Pedicles

Pedicles project posterolaterally connecting the body to the neural arch forming the spinal canal.

Articular pillars

- Pars interarticularis, superior and inferior articular facets.

Facet joints

- Diarthrodial synovial lined joints.
- Upper lumbar spine is oriented in parasagittal plane.
- Lower lumbar spine shows oblique orientation and appears mushroom shaped on axial imaging.

Laminae and Spinous process

Laminae extend posteriorly from articular pillars to join and form the spinous process which projects inferiorly and posteriorly.

LIGAMENTS

Anterior longitudinal ligament

- ALL extends from basiocciput to S1.

- MRI-ALL appears as a thin low signal structure in both T1 and T2 weighted sequences in direct contact with the ventral surface of vertebral bodies.

Posterior longitudinal Ligament

- PLL extends from C1 to S1.
- MRI shows PLL as a low signal structure in both T1 and T2 weighted sequences that is moulded to the posterior disc surface but spans the posterior surface of the body like a bow string with interposed epidural fat and veins.

Ligamentum Flavum

- Ligamentum flavum extends from the anterior aspect of the lower margin of one lamina to the posterior surface of the lamina below. Axial imaging shows V shaped structure.

SOFT TISSUE COMPONENTS

Epidural Fat and Veins

Epidural fat surrounds the thecal sac and root sleeves and appears high signal on T1 weighted sequences. Epidural venous plexus lie between Posterior longitudinal Ligament and posterior surface of vertebral body. Basivertebral veins drain into the plexus and appear as low signal voids in MRI that shows enhancement on contrast administration.

NEURAL TISSUES

Spinal Cord

Spinal cord extends from Cervicomedullary junction to L1-2. Conus medullaris is the diamond shaped terminal part of cord at L1-2. Cauda equina consists of nerve roots from conus appearing like horsetail. Filum terminale is the pial extension of meninges which shows crescent shaped appearance (axial imaging) surrounded by the lower Sacral roots dorsally and Lumbar roots anterolaterally.

Lumbar Nerve Roots and Neural Foramina

- *Nerve roots* exit the spinal canal at 45^{o} and carry a sleeve of dura called Axillae.
- Motor nerve roots lie ventral to sensory nerve roots.

Neural Foramina and Dimensions

Boundaries

- **Anterior-**Vertebral body superiorly, disc and, Posterior Longitudinal Ligament inferiorly.
- **Posterior-**Ligamentum flavum and articular facet.
- **Superior and Inferior-**Pedicles

Superior part of neural foramen is widest through, which the nerve root exits.

Sacral Plexus

Ventral rami of L4-5 and S1-4 nerves forms the sacral plexus

THORACIC SPINE

ANTERIOR ELEMENTS

- ***Shape***-Cone Or Triangular. Slightly wedged shaped from front to back.
- ***Intervertebral Discs***-Height less than cervical or lumbar discs but annulus is thicker.

POSTERIOR ELEMENTS

Pedicles and Laminae

Pedicles project posteriorly from the superior aspect of body. Laminae are broad and short and overlap like the tiles on a roof. Spinous process is long and gracile project posteriorly and inferiorly.

Articullar Pillars and Joints

Facet joint lie in the coronal plane. Transverse process project laterally. T1-T10 transverse process articulates with first 10 ribs (Costotransverse joints).

LIGAMENTS

Anterior Longitudinal Ligament and Posterior Longitudinal ligaments are thicker.

NERVES AND NEURAL FORAMINA

Spinal cord gives rise to a small ventral and large dorsal nerve root that exits through the neural foramina.

CERVICAL SPINE

C1-Bony ring with ellipsoid superior facets articulates with the occipital condyles to form the atlantooccipital joint. The inferior facets articulate with superior facets of C2 to form the atlantoaxial joint. C2 has dens, which projects superiorly nearly up to the

clivus. Dens articulate with the anterior arch of C1. C3-C7 Vertebrae are functionally and anatomically similar.

ANTERIOR ELEMENTS

Vertebral and Uncovertebral joints

Cervical vertebrae are box shaped. There is a Increase in size from C3-7. Superior uncinate process articulates with the vertebral bodies above to form the uncovertebral joints.

Intervertebral discs

Bean shaped with concave posterior margin. Nucleous pulposus appears high signal on T2 weighted sequence and Annulus fibrosis appears low signal on T1 and T2 weighted sequences.

POSTERIOR ELEMENTS

Pedicles

Short and project posteriorly and laterally.

Articular facets

Laminae and spinous process

Laminae are thin plates that fuse in the mid line to form the spinous process, which is often bifid. C7 has the longest spinous process.

NEURAL FORAMINA AND NERVES

Boundaries

- **Anterior** –Vertebral body.
- **Posterior**-Articular pillars and Ligamentum Flavum.
- **Superior and inferior**-Pedicles

Nerves

The nerves exits at 45^{o} and lies in the Inferior half of the neural foramina with superior half occupied by fat and epidural veins. The Dorsal roots lie behind and above the ventral roots.

LIGAMENTS

The various ligaments include Anterior longitudinal ligament, Posterior longitudinal ligament and Ligamentum flavum. The union of Superior and Inferior cruciate ligaments behind the dens forms Transverse Ligament.

Epidural fat and Veins

Fat is sparse. Epidural veins are larger.

SPINAL CORD

Shape on axial imaging

Cervical cord appears elliptical while the thoracic cord appears round. Conus medullaris is the diamond shaped expansion of cord which terminates at L1-2. Filum terminale shows crescent shaped appearance.

Fissures

Ventral median fissure, Dorsal median sulcus, Dorsal lateral sulci, Dorsal intermediate sulci.

Central grey matter-H shaped with Anterior and Posterior Horns.

White matter-Anterior, lateral and dorsal funiculi.

MENINGES

Dura and subdural space

Spinal dura is continuous with the cranial dura and extends upto S2 where it blends with filum terminale and encloses the subdural space, which is usually small.

Arachnoid and subarachnoid space

Arachnoid is loosely attached to the dura and encloses the subarachnoid space.

Subarachnoid space is widest at crainovertebral junction.

AP dimensions of spinal canal

- **C3-C7**-10-15mm.
- **D1-D12**-12-13mm.
- **L1-L5**-15-20mm.

GENERAL EMBRYOLOGY

Ovum+Sperm

↓

Fertilized Ovum

↓

2cel → 3cell → 4Cell stages

↓

16cell stage (Morula)

↓

Blastocyst

↓

Bilaminar Embryonic Disc with amnionic cavity and
yolk sac surrounded by Trophoblast

PROCHORDAL PLATE is a Circular area in endoderm due to change of cubical cells of endoderm to columnar cells. It determines the central axis and the head end of the embryo.

PRIMITIVE STREAK is a Proliferation of the ectoderm along the central axis in the tail end of embryo. These cells spread from the primitive streak between the ectoderm and endoderm to form the intraembryonic mesoderm. The mesoderm fails to separate the ectoderm and endoderm in the region of prochordal plate and cloacal membrane.

FORMATION OF NOTOCHORD

Notochord extends from the cranial end of primitive streak to the caudal end of prochordal plate and most of it disappears with remnants persisting as Nucleus pulposis.

NEURAL TUBE FORMATION

The ectoderm destined to develop into neural tube lies dorsal to the notochord called neural plate. Dorsal groove formed called neural groove becomes deeper until the two edges of neural plate come close to each other and fuse thus converting the neural groove into neural tube. The enlarged cranial part forms the brain while the caudal part forms the spinal cord.

DEVELOPMENT OF SPINAL CORD

Caudal part of neural tube gives rise to the spinal cord. It has 3 layers namely ependymal layer, mantle layer and marginal layer. Mantle layer differentiates into ventral basal lamina and dorsal alar lamina which gives rise to the anterior horn, posterior horn and roots respectively. The marginal layer gives rise to the ascending and descending tracts. The ependymal layer lines the central canal of the spinal cord and extends the entire length of vertebral column. Later the vertebral column grows faster than the spinal cord resulting in recession of spinal cord causing the spinal roots to course obliquely to reach their respective neural foramina.

FORMATION OF DISTAL CORD

The distal spinal cord forms by the processes of canalization and retrogressive differentiation. After neurulation is complete on Day 26-27 the caudal end of the neural tube and the caudal end of the notochord blend into a large aggregate of undifferentiated cells designated the caudal cell mass. The caudal cell mass extends into the tail fold, adjacent to the distal end of the developing hindgut and the mesonephros. This juxtaposition of developing genitourinary, Notochordal, and Neural structures within the tail fold appears to account for the common concurrence of distal vertebral, Neural, Anorectal, Renal, and Genital anomalies.

Within the caudal cell mass, small vacuoles form, coalesce, and eventually connect with the central canal of the spinal cord above, "canalizing" the caudal cell mass. A

focal widening of this canal at the distal conus or proximal filum is designated the terminal ventricle

The most cephalic portion of this distal cord forms the lower half of conus medullaris. Major portion of this distal cord undergoes involution to a glio-ependymal strand called the filum terminale by a process designated retrogressive differentiation.

The primitive filum extends caudally from the apex of the primitive conus to the 29th-30th vertebra. After involution of the distal cord to the filum terminale, the newly formed conus medullaris lies opposite the lower coccygeal segments. Thereafter, the spinal cord and filum do not shorten further. Rather, they elongate and thicken by interstitial growth. The caudal cell mass formed by notochord, mesoderm, and neural tissue simply segments into somites to form the sacral, coccygeal, and tail vertebrae. Retrogressive differentiation then leads to reduction of most of these segments with loss of the tail. Thereafter the vertebral column elongates with growth and grows faster than the cord. All further "ascent" of the cord results from disproportionately greater longitudinal growth of the vertebrae, not involution; the bones simply grow faster and descend away from the cord.

FORMATION OF MENINGES

The ventral part of the neural plate induces the surrounding mesenchyme to form the meninges.

FORMATION OF VERTEBRAL COLUMN

The mesoderm on either sides of the notochord is called paraxial mesoderm. It is segmented to form somites.

Somite has 3 parts

- **Sclerotome**-Vertebral column and Ribs.

- **Myotome**-Paraspinal muscles
- **Dermatome**-Skin and subcutaneous tissue.

REVIEW OF LITERATURE

Congenital malformations of the Spine are grouped into three broad categories[1, 7]

SPINA BIFIDA APERTA

Lesions in which the neural tissue is exposed to view in the midline of the back are designated spina bifida aperta. The most common forms are the Myelocele and Myelomeningocele; Meningoceles without skin cover are less common.

OCCULT SPINAL DYSRAPHISM

Lesions in which the neural tissue lies deep to an intact skin cover are designated occult spinal dysraphism[5, 19.] and includes Dorsal dermal sinus, Spinal lipoma, Tight Filum Terminale syndrome, Neurenteric cyst, and Diastematomyelia. Midline cutaneous stigmata such as skin dimples, hemangiomas, and hypertrichosis frequently overlie the zone of abnormal nervous tissue.

CAUDAL SPINAL ANOMALIES

Malformations of the distal end of the spine, spinal cord, and meninges are associated with disorders of the hindgut, kidneys, urinary bladder, and genitalia. These include Sacral agenesis, Terminal myelocystocele, and Anterior sacral meningocele ect.

INCIDENCE

- 2-4/1000 live births in India.

GENDER DISTRIBUTION

The prevalence rate of Myelomeningocele[3] Spinal lipomas and Diastematomyelia is slightly higher in girls (Ratio of 1.2:1). Meningocele, Dorsal dermal sinus, Thick filum terminale syndrome have no gender predilection.

AGE OF PRESENTATION

Open Spinal dysraphism[22, 24] presents at birth, but spina bifida occulta may not be obvious until later in childhood or early adult life[32].

SPINA BIFIDA APERTA

Definition

Form of spinal dysraphism in which the neural tissue and/or meninges are exposed to the environment, because the skin, fascia, muscle, and bone are deficient in the midline of the back. Myelocele and Myelomeningocele[7] are the two commonest forms of spina bifida aperta.

Incidence

Myelocele and Myelomeningocele occur in 1-2 persons per 1, 000 live births.

Gender Distribution

Myelomeningocele afflicts females more commonly than males.

Distribution in spinal cord

Thoracic 2%, Thoracolumbar 32%, Lumbar 22%, and Lumbosacral 44%[1]

Clinical features

- Sensorimotor deficits of the lower extremities
- Incontinence of bladder and bowel.

Embryology

If the neural folds fail to flex and to fuse into a tube, they persist instead as a flat plate of neural tissue. This flat plate of unneurulated neural tissue is designated the neural placode. Because the neural tube does not close, the superficial ectoderm cannot disjoin from the neural ectoderm and remains in lateral position. Therefore the skin that develops from the ectoderm also lies lateral in position, leaving a midline defect. Mesenchyme then cannot migrate behind the neural tube, so the bony, cartilaginous, muscular, and ligamentous elements are also deficient in the midline. Instead, the bones, cartilage, muscle, and ligament develop in an abnormal position ventral-lateral to the neural tissue, and appear bifid and "everted. " The unfused neural plate is thus exposed to view in the midline of the back at the site of the midline deficiency of skin, bone, cartilage, muscle and ligament.

Local Examination

The neural tissue appears as a raw, reddish, vascular oval plate.

Relation of Meninges to neural Tissue

The pia-arachnoid membrane covers the ventral surface of the neural plate.

Relation of dorsal and ventral nerve roots to neural tissue

The two ventral motor nerve roots arise from the ventral surface of the neural plate just to each side of the midline ventral sulcus. The paired dorsal sensory roots arise lateral to the ventral roots.

Role of imaging

Rarely untreated Myelomeningocele comes for imaging, as the pathology is clearly visible and taken for surgical repair in the neonatal period. Most commonly repaired lesions come for imaging for follow up and assessment of retethering of cord.

Associated anomalies

- ***Spinal Curvature-*** Scoliosis, Lordosis, Kyphosis.
- ***(Epi) dermal inclusion cysts***
- ***Lipomas*** -6%
- ***Hydromyelia-****30% to 50%*
- ***Diastematomyelia-***31%to46%with Myelomeningocele(complex spina bifida)[21].
- ***Chiari II Malformation-*** Nearly always associated.

OCCULT SPINAL DYSRAPHISM

MENINGOCELE

Definition-Dorsal protrusion of meninges and CSF without neural elements with skin covering.

Etiology-Acquired more common than congenital.

Incidence-1per 10,000 live births.

Gender-Equal.

Location- 80% situated in lumbosacral spine

Clinical Features

- Lumbosacral mass.
- Neurological deficits uncommon.

Imaging

Plain Radiograph –Soft tissue swelling and Spina bifida.

(CT/MR)- Dorsal protrusion of meninges and CSF without neural elements.

THE DORSAL DERMAL SINUS

Definition

Mid line epithelial lined tract extending from skin to a variable extent into the meninges and neural tissue of spinal cord[10].

Incidence: Uncommon

Gender: No gender predilection

Embryology

If the superficial ectoderm fails to separate from the neural ectoderm at one point, a focal segmental adhesion is formed. Different rates of growth between neural and spinal tissue lead to "ascent" of the cord, the local adhesion is drawn out into an elongated epithelial-lined tube that still connects the spinal cord with the skin of the dorsal surface called Dorsal dermal sinus.

Location

Lumbosacral-57% and Occipital-24% (Wright's series[10]).

Clinical features

Symptoms of infection of sinus tract or mass effect of associated epidermoid/dermoid.

Local Examination

The sinus frequently appears as a pinpoint hole or a small atrophic zone in the skin typically midline, with hemangiomas and tuft of hair.

Imaging

- **CT-**Relatively hyperdense tract that traverses the skin, subcutaneous tissue ,dysraphic spine and ends variably at the dura, sub arachnoid space, spinal cord or ends in Dermoid or Epidermoid.

- **MRI**- best delineates the extent and direction of tract ***Associated anomalies***

- Epidermoids and Dermoids (60%)
- Tethering of cord (80%)
- Lipoma (15 to 20%)

Differential Diagnosis

- **Pilonidal Sinus** lie in low location, near to anus, and extend inferiorly or horizontally toward or to the dorsal surface of the coccyx. It does not enter the spinal canal. (*Haworth JC etal*[70]*)*

- **Sacral dimples** may occur together with spinal dysraphism.

DERMOID AND EPIDERMOID TUMORS

Definition-Epidermoid cysts are lined by a membrane composed only of superficial (epidermal) elements of the skin[25]. Dermoid cysts are unilocular or multilocular. These tumors may arise as congenital rests or as iatrogenic implantation of viable skin elements during surgery or during spinal taps. Approximately 25% of (epi)dermoid tumors form in association with dermal sinuses[28].

Distribution-Spinal Dermoid and Epidermoid tumors constitute about 15% of all CNS (Epi)dermoids. Dermoid and Epidermoid tumors occur equally. In Lunardi's series[71], Epidermoids were most frequently in Lumbar region; Dermoids were most frequently dorsal (25%) or dorsolumbar (75%). 38% were intramedullary and 63% intradural extramedullary.

Crossectional imaging-Epidermoid tumors are usually isointense with CSF or just slightly hyperintense to CSF. So they may be difficult to discern within the spinal canal. Fat-containing portions of Dermoids will manifest as lesions with high signal on T1 series and low signal on T2 series.

SPINAL LIPOMA

Definition

Distinct collections of fat and connective tissue that are partially encapsulated and have a definite connection with the spinal cord or leptomeninges

Incidence

Spinal lipomas are the most common type of occult spinal dysraphism and account for 35% of skin-covered Lumbosacral masses.

Gender

More common in females, with a sex ratio of about 1. 5:1 to 2:1.

Principal categories

Lipomyelomeningocele (84%), Intradural lipoma (4%), Filar lipoma (12%).

Clinical features

The chief complaints are usually a mass on the back (59%), urinary incontinence (23%), or weak/deformed lower extremity, perhaps with trophic ulceration (11%). Cutaneous stigmata include subcutaneous mass, Skin dimple, Hemangioma, hairy-nevus, denuded skin patch, Scar-like patch.

Embryology of lipomyelomeningocele and Intrdural lipomas

Spinal lipomas result from focal premature disjunction of cutaneous ectoderm from neural ectoderm[72]. If the superficial ectoderm separates from the neural ectoderm, before the neural tube has closed completely, then mesenchyme may gain access to the interior of the closing neural tube. Such mesenchyme could prevent closure of the neural tube focally, leading to dorsal myeloschisis. The mesenchyme is induced by the dorsal surface to form fat.

The mesenchyme surrounding the ventral surface of the closing plate, i.e, the future exterior of the neural tube is induced by the ventral surface to form meninges. No meninges would form in the midline dorsally since there is no ventral surface to induce them there. This would leave a midline dorsal defect in the meninges. Similarly, improper neurulation would prevent proper development of the neural arches, fascia, and muscle, creating a posterior spina bifida. The fat induced by the dorsal surface of the neural plate could then extend directly posteriorly, through the gap in the meninges and through the spina bifida into the subcutaneous tissue of the back. The fat would be anatomically extradural. The junction of fat and meninges would necessarily lie at the neural ridge that divides the dorsal and ventral surfaces of the neural folds.

3 Groups: Lipomas with intact dura, Deficient dura, and Filar lipomas.

Spinal Lipomas with Intact Dura (Intradural lipomas)

The intradural lipomas[20] appear to arise in the dorsal midline of a cleft spinal cord and then bulge outward to form subpial masses of fat. They affect the cervical and thoracic spinal cords predominantly. In this group the spinal canal is usually nearly normal with narrow spina bifida or focal segmentation anomalies. The canal itself may be expanded by the mass. The dura is thinned, perhaps translucent, but remains intact and is displaced peripherally by the combined mass of cord and lipoma. The lipoma typically lies dorsal or dorsolateral to the cord; frequently causes cord rotation and frequently cause high-grade stenosis or block.

Imaging

- **CT-**Fat atteunation intradural mass
- **MRI-**High signal intradural mass in dorsal position in T1Weighted images

Spinal Lipomas with Deficient Dura (lipomyelocele and lipomyelomeningocele)

The most common forms of spinal lipoma (84%) are associated with definite defects in the dura through which the lipoma may extend from the spinal cord to the subcutaneous tissue. The subcutaneous component of the mass typically forms a lump in the low back. The subjacent spinal canal shows a wide spina bifida. Segmentation anomalies are present in 50%. The spinal cord beneath the Lipoma is cleft dorsally and very closely resembles the neural plate of a Myelomeningocele. The dura is deficient in the dorsal midline, deep to the lipoma. . Lipomyelocele and lipomyelomeningocele are exactly analogous to the Myelocele and the Myelomeningocele, with the fat inserting into the dorsal surface of the neural plate and closure of the skin over the fat.

Imaging

- **CT**-Fat atteunation mass attached to neural placode
- **MRI**-High signal mass attached to the cord with tethering in T1weighted images.

Filar Lipomas

Imaging –High signal mass attached to filum terminale in T1weighted images.

Associated Anomalies

Dermal sinuses(20%), Teratomas(3%-7%), Diastematomyelia(1%-6%), Hydromyelia (2.5% -24%), Chiari I malformation(1%-3%), Genito-urinary lesions (4%) and Genito-anal lesions (4%).

DERANGED CANALIZATION AND RETROGRESSIVE DIFFERENTIATION

TIGHT FILUM TERMINALE SYNDROME

Definition

In this syndrome there is traction on the spinal cord as a result of an abnormally short and thick filum terminale[4].

Criteria

- The filum must measure > 2 mm in diameter
- No other cause for tethering present. The tip of the conus medullaris lies below L2 in 86%.

Embryology

Failure of complete involution of the distal cord (Raghaven N etal[6])

Age; 3-35 yrs.

Gender; No predilection.

Clinical features

Sesorimotor Deficits and Bowel And Bladder Disturbances.

Imaging

- **Plain radiograph and CT**-Dysraphic spine, vertebral segmentation anomalies.
- **Myelography (Conventional and CT)** –Shows low lying conus and thick filum. Exiting nerve roots have a lateral or uphill course.
- **MRI**- Low lying conus and thick filum with associated lipoma.

Associated anomalies

Filar fibrolipomas 29%, Kyphoscoliosis 15% to 25%.

SACROCOCCYGEAL TERATOMAS

Sacrococcygeal teratoma[26] is a congenital tumor of the caudal pole of the body tissues derived from all three germ layers It is the most common newborn tumor, the most common tumor of the sacrococcygeal region in childhood.

Etiology

Sacrococcygeal teratomas most probably arise from totipotential cells derived from Hensen's node.

Incidence

1 per 35,000-40,000 births.

Gender

Females predominate (80%). Hereditary forms have been reported.

Presentation

Patients may present asymptomatically because of an external mass or they may manifest hydrops, high output cardiac failure, respiratory failure, and renal insufficiency related to the bulk and vascularity of the mass

Altman Classification

Type I tumors (47%) have predominantly external component with minimal presacral component.

Type II tumors (35%) are evident externally but have significant intrapelvic extension.

Type III tumors (9%) can be detected externally but lie predominantly within the pelvis and abdomen.

Type IV tumors (10%) are entirely presacral.

Crossectional Imaging-Most sacrococcygeal tumors are mixed solid and cystic lesions that form a large mass caudal to the coccyx. Five percent are predominantly cystic. Calcification is found in about 50% of benign teratomas.

TERMINAL VENTRICLE

The terminal ventricle is the normal slight expansion of the central canal of the cord within the distal conus and/or proximal filum. It appears to represent the point of union between the portion of the central canal made by neurulation and the portion made by canalization of the caudal cell mass. MRI studies often demonstrate a tiny drop of CSF at the site of the terminal ventricle. Slight expansion of this in patients without other pathology is presently regarded as normal.

TERMINAL SYRINGOHYDROMYELIA

Large cystic expansion of the distal one-third of the cord is found alone or in association with diverse forms of occult spinal dysraphism. Terminal hydrosyringomyelia[57] may be seen by MRI in up to 30% of patients with occult spinal dysraphism. It is found most frequently in patients with concurrent anorectal anomalies (67%), Meningocele (54%), Diastematomyelia (38%), Tight filum terminale syndrome (33%), Lipomyelomeningocele (19%), and Dermal sinus tract (17%)

CAUDAL SPINAL ANOMALIES WITH ANORECTAL AND UROGENITAL MALFORMATIONS

Embryogenesis

Union of the hindgut, allantois, and wolffian duct forms a common cloaca just ventral to the notochord and near to the caudal cell mass. Appearance of a urorectal septum then divides the cloaca into a dorsal hindgut and a ventral urogenital sinus. At 7 weeks' gestation the cloacal membrane is bisected to form the anal membrane and the urogenital membranes. High and low imperforate anus, rectovaginal fistula and bladder, cloacal exstrophy, persistent cloaca, and genital anomalies all result from maldevelopment of the cloaca, urorectal septum, anal membrane, and urogenital membrane.

TERMINAL MYELOCYSTOCELE (SYRINGOCELE)

Terminal myelocystocele is a cystic dilatation of the distal spinal cord. Terminal myelocystoceles constitute 1% to 5% of skin-covered lumbosacral masses. They are typically associated with the OEIS and other severe anomalies of the hindgut and genitourinary systems. It may also be related to the teratogen retinoic acid.

SYNDROME OF CAUDAL REGRESSION

The syndrome of caudal regression[12] designates a constellation of anomalies of the hind end of the trunk, including partial agenesis of the thoracolumbosacral spine, imperforate anus, malformed genitalia, bilateral renal dysplasia or aplasia, pulmonary hypoplasia, and, in the most severe deformities, extreme external rotation and fusion of the lower extremities (Sirenomelia).

Incidence

Sacral Agenesis[14] occurs in approximately 1 per 7, 500 births.

Gender

Males and females are affected equally. Nearly all cases are sporadic.

Diabetes Association

There is a definite but incomplete association with Diabetes mellitus.

Clinical Features

Sensory/motor deficits and bowel/bladder disturbances.

Imaging

- **PLAIN RADIOGRAPH**-shows variable absence of distal spine.
- **CT/MRI** best characterizes the caudal spinal anomalies.

ANTERIOR SACRAL MENINGOCELES

Definition

Anterior sacral meningoceles are diverticulae of the thecal sac that protrude anteriorly into the extra peritoneal presacral space[15].

Inheritance

Most anterior sacral meningoceles[18, 50] occur sporadically. They may be seen in conditions with dural ectasia such as Neurofibromatosis and Marfan's syndrome, or may be associated with the Currarino triad of anorectal malformations, sacral defects, and presacral masses.

Incidence

Clinically, anterior sacral meningoceles account for 3.7% of retrorectal tumors[48.]

Gender

In adults, females appear to be more affected (approximately 10:6). The lesion is equally frequent in boys and girls below age 15,

Clinical Features

Local pressure on pelvic organs causes unremitting constipation, urinary frequency and incontinence, dysmenorrhea, dyspareunia, and back pain. Pressure on the nerve roots causes sciatica, diminished rectal and detrusor tone, numbness and paresthesias in the lower sacral region.

Imaging

In anterior sacral meningocele the spinal canal is usually widened with smoothly scalloped margins. The defect in the sacrum is typically asymmetrical. The typical smooth curvilinear defect designated the "**Scimitar Sacrum**" often starts as a partial

unilateral sacral agenesis that becomes remodeled around the hernia ostium with time. In approximately 20% the sacral defect is midline. The sacral dural sac is often widened and patulous. The stalk is typically narrow. The meningocele sac may be unilocular or multilocular.

LATERAL LUMBAR AND THORACIC MENINGOCELES

These lesions are characterized by CSF-filled protrusions of dura and arachnoid through one or several enlarged neural foramina into the paraspinal extrapleural and retroperitoneal tissue. They may be unilateral or bilateral, and are commonly associated with scoliosis. Lateral meningoceles [47]are most common in patients with mesenchymal disorders such as Neurofibromatosis (85%), Marfan's and Ehler-Danlos syndromes.

DERANGEMENTS IN THE NOTOCHORD (SPLIT NOTOCHORD SYNDROME)

Persistence of a midline adhesion between ectoderm and endoderm could cause derangement in the migration of notochordal cells with consequent deflection or splitting of the notochord. This appears to be the genesis of Dorsal enteric fistula, Neurenteric cyst, and Diastematomyelia.

DORSAL ENTERIC FISTULA

Persistence of a patent notochord canal (canal of Kovalevsky) would create a patent fistula from the mesenteric surface of gut through the mesentery and prevertebral tissue, through the vertebral bodies, spinal canal, and spinal cord, and through bifid laminae to an ostium in the midline skin of the back. Such a complete communication is designated the dorsal entericfistula[44].

NEURENTERIC CYST

Definition

Neurenteric cysts[30, 45] are enteric-lined cysts that present within the spinal canal and exhibit a definite connection with the spinal cord and/or vertebrae. They may communicate with an extra spinal component of cyst in the mesentery or mediastinum around a hemi vertebra or through a butterfly vertebra and/or they may attach by a fibrous stalk to the vertebra, mesentery, or gut.

Etiology

Failure of foregut notochord separation during embryogenesis.

Incidence It is exceedingly rare.

Age and Gender

1st or 2nd decade. Slight male Predominance.

Distribution

Thoracic spine-42%. Cervical spine-32%. Posterior fossa-13%. CVJ-10%. Lumbar-rare.

Imaging

- **Myelography** –Intrdural extramedullary mass anterior to spinal cord.
- **CT**-CSF density mass protruding from the spinal canal into the sacrum.
- **MRI** – T1-iso to slightly hyperintense to CSF.
  T2- slightly hyperintense to CSF.

Associated Anomalies-Vertebral anomalies-43%

Differential Diagnosis

Arachnoid cyst, Epidermoid cyst and Inflammatory cysts.

DIASTEMATOMYELIA

Definition

Diastematomyelia signifies a sagittal clefting of the spinal cord, conus medullaris, and/or filum terminale into two, not necessarily symmetrical, hemi cords.

Embryology

Adhesions between the
embryonic ectoderm and endoderm[40]

↓

Accessory neuroenteric canal

↓

Endomesenchymal tract condenses around
the accessory neuroenteric canal

↓

Split spinal cord

Incidence

Uncommon

Gender

Female predominance

Age

3 age ranges: 0-2 years (20%), 4-8 years (40%), and 12-13 years (17%)[38].

Clinical Features

In children, the common presenting complaints[39] are (i) Musculoskeletal deformities (98%) and Pes cavus, (ii) Neurologic deficits (84%) and Incontinence of bladder and bowel, and (iii) Scoliosis (79%) and in adults, Sensorimotor changes (69%) and pain (58%) . Cutaneous stigmata like hair patches, Nevi, Lipomas are common.

Location

Lumbar or lumbosacral spine 45%, Thoracic spine 31%, Thoracolumbar spine 12%. Cervical spine 7% and the sacrum in 1%. "Double" Diastematomyelia occurs in < 1%. In Diastematomyelia, [16, 17] the conus medullaris is usually low in position. The two Hemi cords nearly always (91%) reunite distally into a reformed cord below the cleft. Hemi cords are asymmetrical in 30%. The filum terminale is thickened and tether the reunited cord.

SPLIT CORD MALFORMATION [5]

- **Type 1-**Two hemi cords separated by osteocartilagenous septum and contained in separate dural sacs.
- **Type11-**Two hemi cords separated by fibrous septum and contained common dural sac.

Imaging

- **PLAIN FILM and NECT** shows the bony septum and associated vertebral anomalies.
- **MYELOGRAPHY-**Depicts the hemi cords.
- **MRI**- Depicts the hemi cords, nature of septum especially fibrous septum and associated anomalies[43].

Associated Anomalies

Segmentation anomalies (85%), Scoliosis and kyphosis(50% - 60%), Klippel-feil syndrome(2%-7%), Sprengel's deformity (7%), Chiari I (3%) Hydrosyringomyelia (50%), Dermal sinus (3%), Lipomyelomeningocele (3%) Teratomas(3%)and Horseshoe kidneys.

SEGMENTATION ANOMALIES OF VERTEBRAL BODIES

Embryology

During the 9-10th week of gestation two ossification centres develop for the ventral and dorsal half of the vertebral body.

Anomalies

- **Asomia**-Complete absence of vertebral body.
- **Hemivertebra**
    - Unilateral wedge vertebra (right or left)-Scoliosis at birth.
    - Dorsal hemi vertebra-Rapidly progressive scoliosis.
    - Ventral hemivertebra(very rare).
- ***Coronal cleft***

Failure of fusion of anterior and posterior ossification centres

- **Butterfly vertebra**

Failure of fusion of lateral halves due to persistence of notochordal tissue.

- ***Block vertebra***-congenital vertebral fusion.

MATERIAL AND METHODS

- The study comprises of 70 patients including 33 males and 37 females, age ranging from birth to 30 years. The study was conducted for a period of 20 months from January 2005 to August 2006 in Barnard Institute of Radiology, Madras Medical College, Chennai.

- The patients were referred from Institute of Child Health, Egmore and Institute of Neurology, Government General Hospital, Chennai to Barnard Institute of Radiology for radiological evaluation.

- Clinically the most common cause for referral was swelling in the back predominantly Lumbosacral region. The other symptoms were sensory/motor deficit, bladder/ bowel disturbances, spinal curvature deformities, cutaneous features like dermal dimple, hypertrichosis, silky hair, dermal sinus and capillary hemangioma ect.

CRITERIA

Inclusion Criteria

- All cases of open spinal dysraphism.
- Cases presenting with lumbosacral swelling.
- Cases presenting with cutaneous stigmata like Dermal dimple, tuft of hair, Nevi, dermal sinus ect.
- Cases showing vertebral anomalies in Plain radiograph.
- Cases presenting with congenital scoliosis/ kyphoscoliosis/ lordosis ect.
- Cases presenting with bladder/bowel incontinence since childhood.

- Cases presenting with motor or sensory deficit since childhood.

Exclusion Criteria

- Treated cases.
- Spinal tumors.

CT TECHNIQUE

All the examinations were performed on a **Toshiba Asteion Spiral CT scanner.**

PARAMETERS

LUMBAR SPINE

- Axial contiguous images from L1 TO S1.
- Slice thickness=4mm.
- Intersection gap=3-4mm.
- Window settings=wide and narrow.
- Sagittal and coronal reconstruction using multiplanar reconstruction technique.

DORSAL SPINE

- Axial contiguous images D1 TO D12.
- Slice thickness=4mm.
- Intersection gap=4mm.
- Window settings=wide and narrow.
- Sagittal and coronal reconstruction using multiplanar reconstruction technique.

CERVICAL SPINE

- Axial contiguous images CVJ TO D1.
- Slice thickness=2-3mm.
- Intersection gap=2mm.
- Window settings=wide and narrow.

- Sagittal and coronal reconstruction using multiplanar reconstruction technique.

MRI TECHNIQUE

MRI was performed using **1.5 Tesla super conducting SIEMENS MAGNETOM , symphony.**

PARAMETERS

LUMBAR SPINE

1. Sagittal fast spin echo, T1-weighted image from conus to S1.

- Field Of View=28cm.
- Slice thickness=4-5mm.
- Intersection gap=1mm.
- Matrix=256x512.

2. Sagittal fast spin echo T2-weighted image.

Field Of View=28cm.

Slice thickness=4-5mm.

Intersection gap=1mm.

Matrix=320x512.

3. Axial fast spin echo T2-weighted image.

- FOV=20cm.
- Slice thickness=4-5mm.
- Intersection gap=1mm.
- Matrix=256X256.
- HASTE myelogram sequence.

DORSAL SPINE

1. Sagittal T1-weighted sequence including the entire Dorsal spine.

- FOV=40cm.
- Slice thickness=3-4mm.
- Intersection gap=1mm.
- Matrix=512x512.

2. Sagittal fast spin echo T2-weighted sequence including the entire dorsal spine

- FOV=40cm.
- Slice thickness=3-4mm
- Intersection gap=1mm.
- Matrix=512x512.

3. Axial T2-weighted sequence including the entire Dorsal spine.

- FOV=40cm.
- Slice thickness=3-4mm.
- Intersection gap=1mm.
- Matrix=512x512.
- HASTE Myelogram.

CERVICAL SPINE

1. Sagittal fast spin echo, T1-weighted image including from the cerebellar tonsils to D1.

- FOV=26-28cm.
- Slice thickness=3-4mm.
- Intersection gap=1mm.
- Matrix=256x512.

2. Sagittal fast spin echo T2-weighted sequence including from the cerebellar tonsils to D1.

- FOV=40cm.
- Slice thickness=3-4mm.
- Intersection gap=1mm.
- Matrix=384x512.

3.Axial 2D GRE covering C1-C7.

- FOV=20cm.
- Slice thickness=3mm.
- Intersection gap=1mm.
- Matrix=384x512.
- HASTE Myelogram.

For interpretation the following Characteristics of Spinal dysraphism were studied and analyzed in these patients.

1. TYPES

Open Spinal Dysraphism

- Myelomeningocele.
- Myelocele.
- Meningocele.

Occult Spinal Dysraphism

- Spinal lipomas.
- Diastematomyelia.
- Dorsal dermal sinus.
- Tight filum terminale syndrome.
- Anterior sacral meningocele.
- Sacral agenesis.

2. CT CHARACTERISTICS

Lesion Attenuation

- Fluid-Menigocele .
- Soft tissue with fluid-Menigomyelocele.
- Soft tissue-Myelocele.
- Fat with soft tissue-Lipomyelocele.
- Fat with soft tissue and fluid-Lipomyelomeningocele.
- Fat-Dural lipomas, Filar lipomas.

Spinal Location

- Lumbosacral.
- Lumbar.
- Dorsal.

- Cervical.

Vertebral Anomalies

- Spina bifida.
- Butterfly vertebra.
- Hemi vertebra.
- Block vertebra.
- Others.

Spinal Curvature anomalies

- Scoliosis
- Kyphosis
- Lordosis

Septum in Diastematomyelia

- Bony
- Fibrous

3. MRI CHARACTERISTICS [2]

Signal Intensities of lesion in T1-W, T2-W and FLAIR SEQUENCES

- CSF Intensity –Meningocele.
- CSF Intensity+Neural tissue-Myelomeningocele.
- Neural tissue-Myelocele.
- Fat intensity+neural tissue-Lipomyelocele.
- Fat intensity+CSF intensity + neural tissue-Lipomyelomeningocele.
- Fat intensity-Intradural lipomas, Filar lipomas

Septum In Diastemetamyelia

- Bony

- Fibrous

Tethering of cord

Vertebral anomalies

Spinal distribution

Spinal curvature anomalies

Chiari association

Hydromyelia

Hydrocephalus

The above mentioned characteristics of spinal dysraphism were analysed using CT and MRI to arrive at radiological diagnosis.

RESULTS AND ANALYSIS

TABLE 1

OPEN SPINAL DYSRAPHISM

S. NO	Type	Number of cases	Percentage	
			(Out of 70)	(Out of 56)
1	Myelomeningocele	53	75.71%	94.64%
2	Myelocele	2	2.86%	3.57%
2	Meningocele	1	1.43%	1.79%
	Total	56	80%	100%

The most common type of spinal dysraphism is open spinal dysraphism accounting for 80% of the total 70 cases. The most common open spinal dysraphism is

Myelomeningocele accounting for 94.6% of the total 56 cases followed by Myelocele and Meningocele.

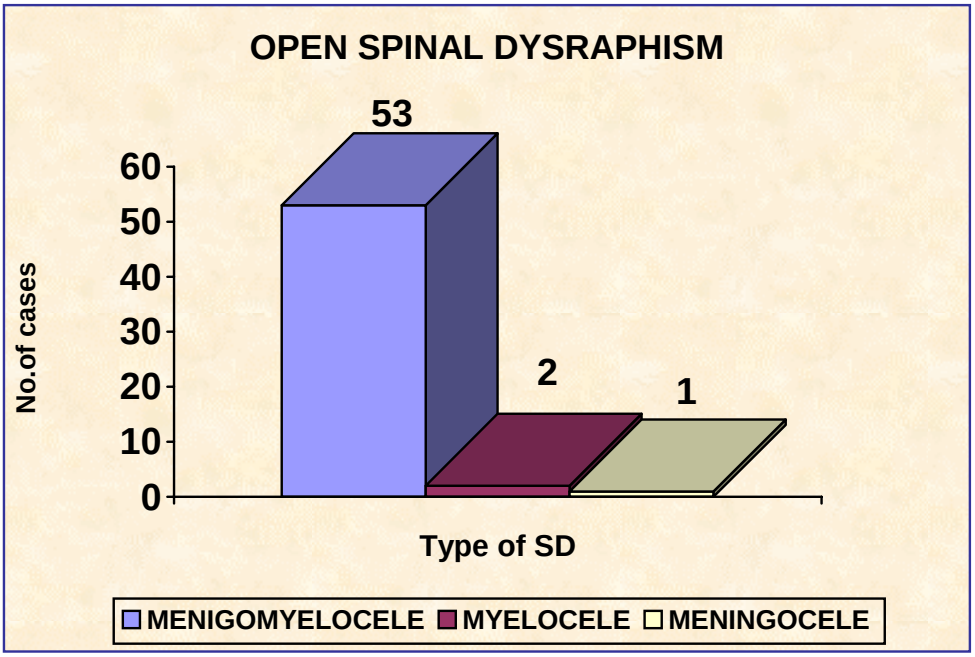

TABLE 2

OCCULT SPINAL DYSRAPHISM

S.No	*Type*	*Number of cases*	*Percentage*	
			(Out of 70)	**(Out of 14)**
1	Spinal lipomas	6	8.57%	42.86%
2	Diastematomyelia	4	5.71%	28.57%
3	Dorsal dermal sinus	1	1.43%	7.14%
4	Tight filum terminale	1	1.43%	7.14%
5	Anterior sacral meningocele	1	1.43%	7.14%
6	Sacral agenesis	1	1.43%	7.14%
	Total	14	20%	100%

Occult spinal dysraphism accounted for 20% of the total 70 cases. The most

common type of occult spinal dysraphism is Spinal lipoma accounting for 42.86% of the total 14 cases.

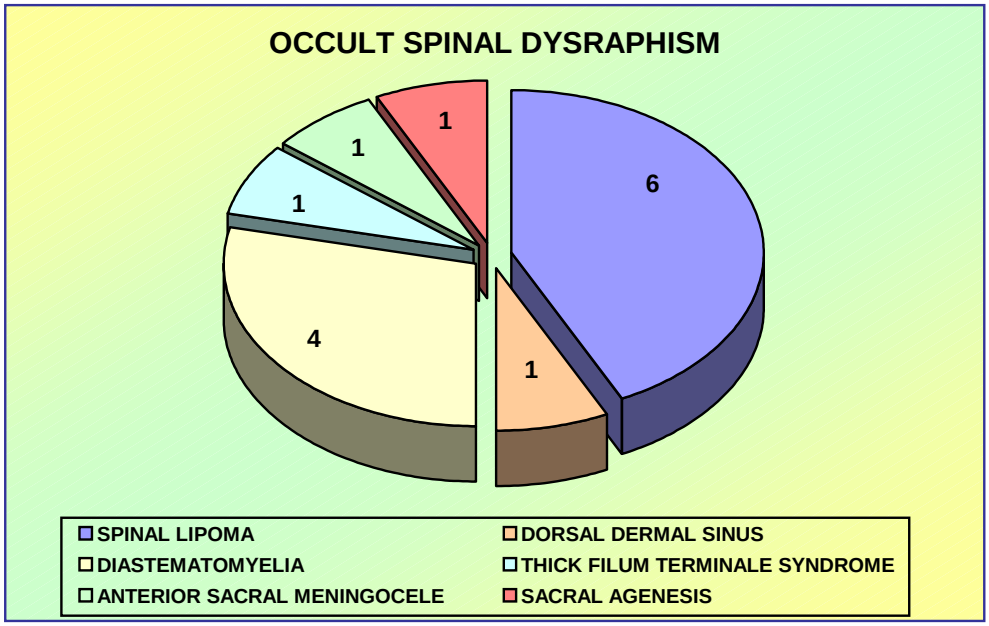

TABLE 3

GENDER DISTIBUTION IN OPEN SPINAL DYSRAPHISM

S.No	*Open spinal dysraphism*	*Number of cases*	*Male*	*%*	*Female*	*%*	*Total*
1.	Meningomyelocele	53	22	41.51	31	58.49	100%
2.	Myelocele	2	1	50	1	50	100%
3.	Meningocele	1	0	0	1	100	100%
	Total	56	23	41.07	33	58.93	100%

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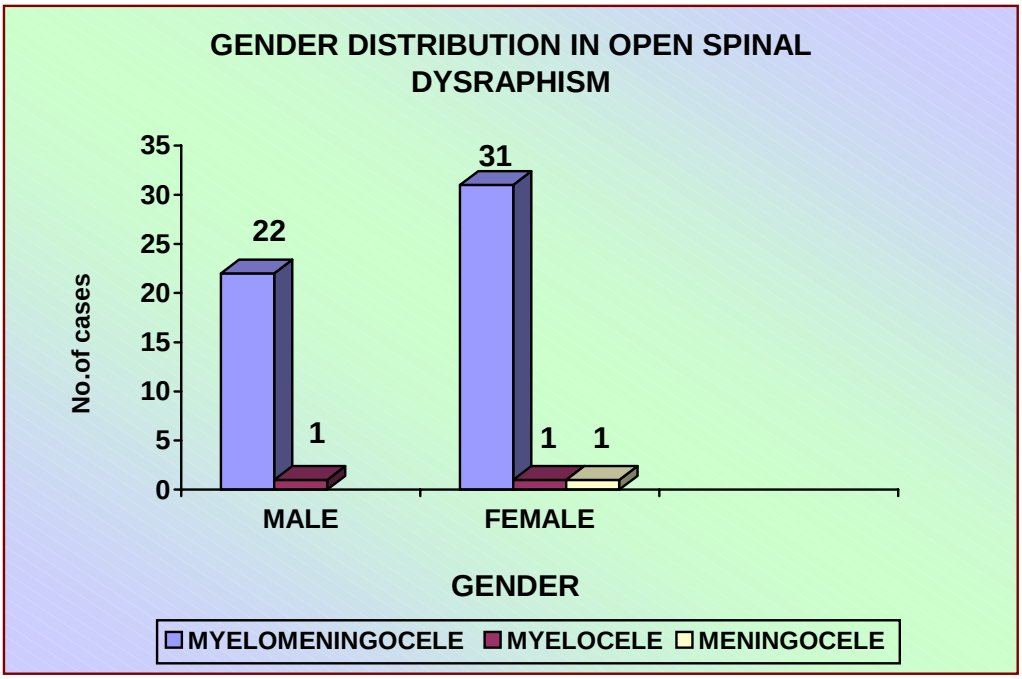
GENDER DISTRIBUTION IN OPEN SPINAL DYSRAPHISM
No.of cases
35
30
25
20
15
10
5
0
22
1
31
1
1
MALE
FEMALE
GENDER
MYELOMENINGOCELE
MYELOCELE
MENINGOCELE

TABLE 4

GENDER DISTRIBUTION IN OCCULT SPINAL DYSRAPHISM

Type	*Number*	*Male*	*%*	*Female*	*%*	*Total*
Spinal lipoma	6	5	83.33	1	16.67	100
Diastematomyelia	4	2	50	2	50	100
Dorsal dermal sinus	1	0	0	1	100	100
Tight filum terminale	1	1	100	0	0	100
Anterior sacral meningocele	1	1	100	0	0	100
Sacral agenesis	1	1	100	0	0	100
Total	14	10	71.43	4	28.57	100

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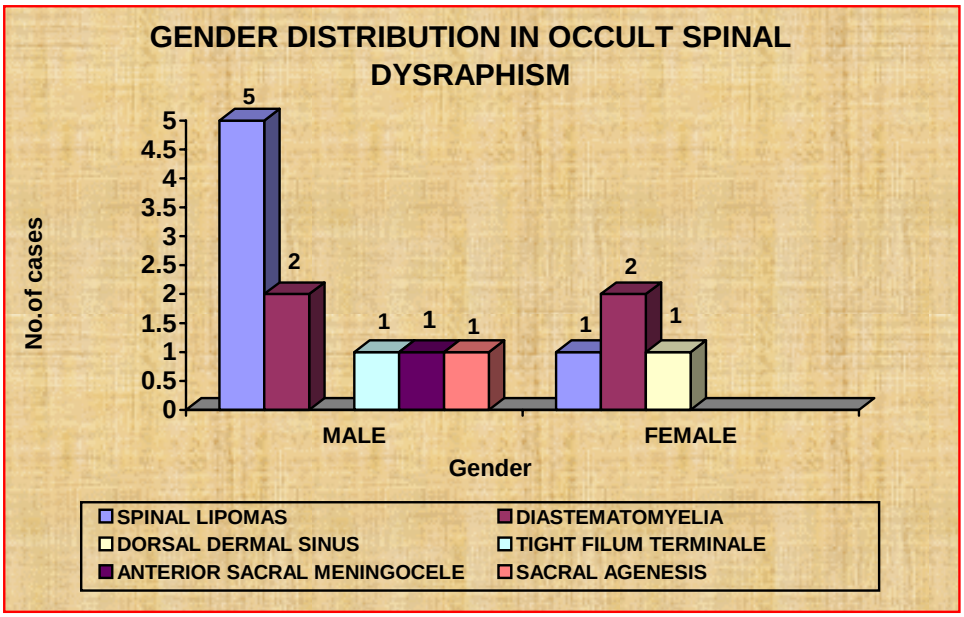

TABLE 5

AGE GROUP DISTRIBUTION IN OPEN SPINAL DYSRAPHISM

S. No	*Age group*	*Myelomeningocele*		*Myelocele*		*Meningocele*		*Total*	
		No of Cases	*%*	*No of Cases*	*%*	*No of Cases*	%	*No of Cases*	%
1	1-10	53	94.64	2	3.57	1	1.79	56	100
2	11-20	0	0	0	0	0	0	0	0
3	21-30	0	0	0	0	0	0	0	0

All cases of open spinal dysraphism occurred in 1 – 10 age group.

Mean age of presentation is 1.23yrs

TABLE 6

AGE GROUP DISTRIBUTION IN OCCULT SPINAL DYSRAPHISM

Age group	**Spinal lipomas**		**Diastema-tomyelia**		**Dorsal dermal sinus**		**Tight filum ternminale**		**Anterior sacral meningocele**		**Sacral agenesis**	
	No	***%***	***No***	***%***	***No***	***%***	***No***	***%***	***No***	***%***	***No***	***%***
1-10	4	66.67	4	100	1	100	1	100	0	0	0	0
11-20	2	33.33	0	0	0	0	0	0	1	100	1	100
21-30	0	0	0	0	0	0	0	0	0	0	0	0
Total	6	100	4	100	1	100	1	100	1	100	1	100

Mean age of presentation is 6.57yrs

TABLE . 7

CUTANEOUS MANIFESTATIONS OF OCCULT SPINAL DYSRAPHISM

Cutaneous Signs	*Dermal dimple*	*Hyper-trichosis*	*Silky hair*	*Palpable mass*	*Dermal sinus*	*Capillary hemangioma*	*Total*
No.of cases	2	2	1	7	1	1	14
%	14.29	14.29	7.14	50	7.14	7.14	100

The most common cutaneous manifestation is palpable mass in the back

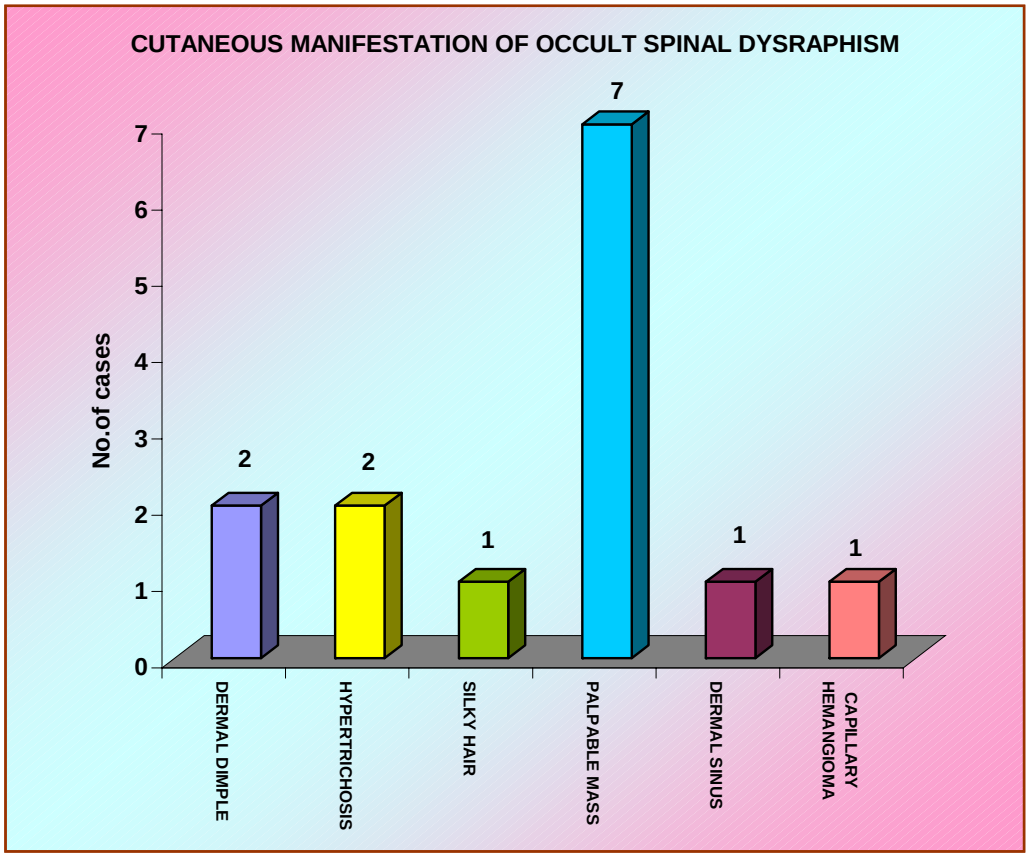

TABLE 8

NEUROLOGICAL MANIFESTATIONS OF SPINAL DYSRAPHISM

Type	*Motor and Sensory deficit*	*Bowel and Bladder incontinence*
Open spinal dysraphism	56	56
Occult spinal dysraphism	6	5

Neurological manifestations occurred in all cases of open SD while in occult SD 11 of the 14 cases showed neurological manifestations.

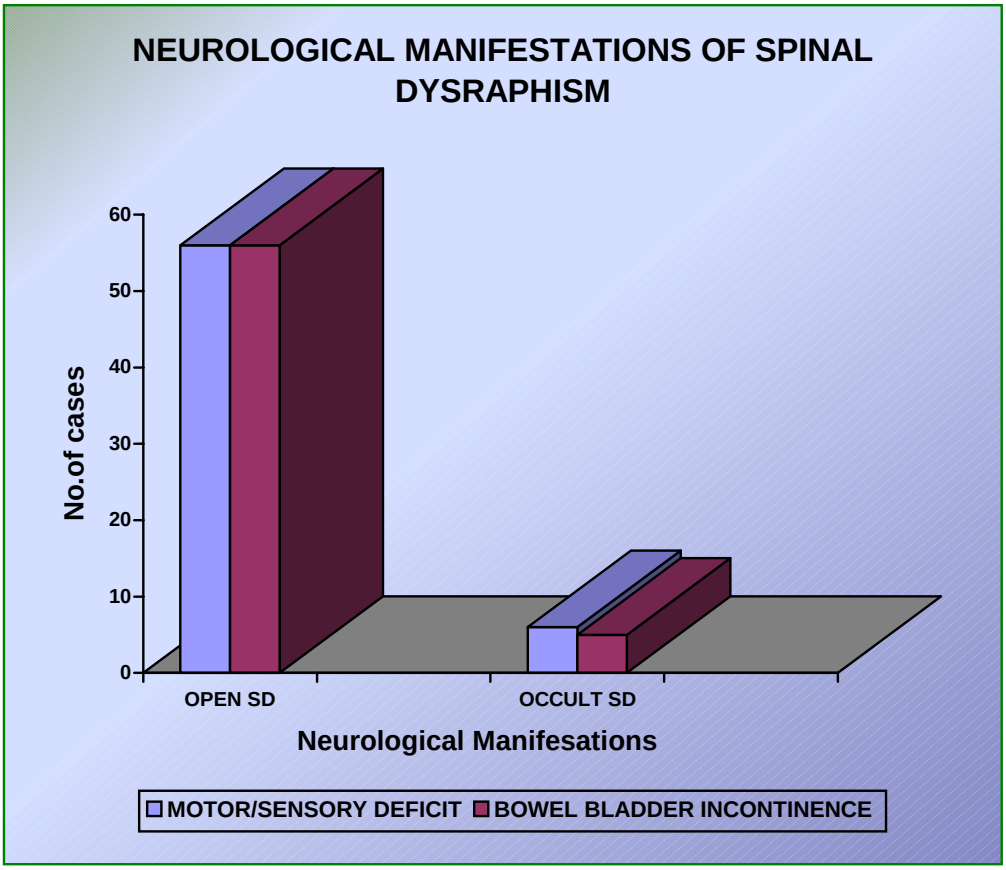

TABLE 9

SPINAL LIPOMAS

Type	*Number of cases*	*Percentage*
Lipomyelocele	1	16.67%
Lipomyelomeningocele	3	50.00%
Dural lipomas	1	16.67%
Filar lipomas	1	16.67%
Total	6	100%

Lipomyelomeningocele is the most common type of spinal lipoma accounting for 50% of the total cases.

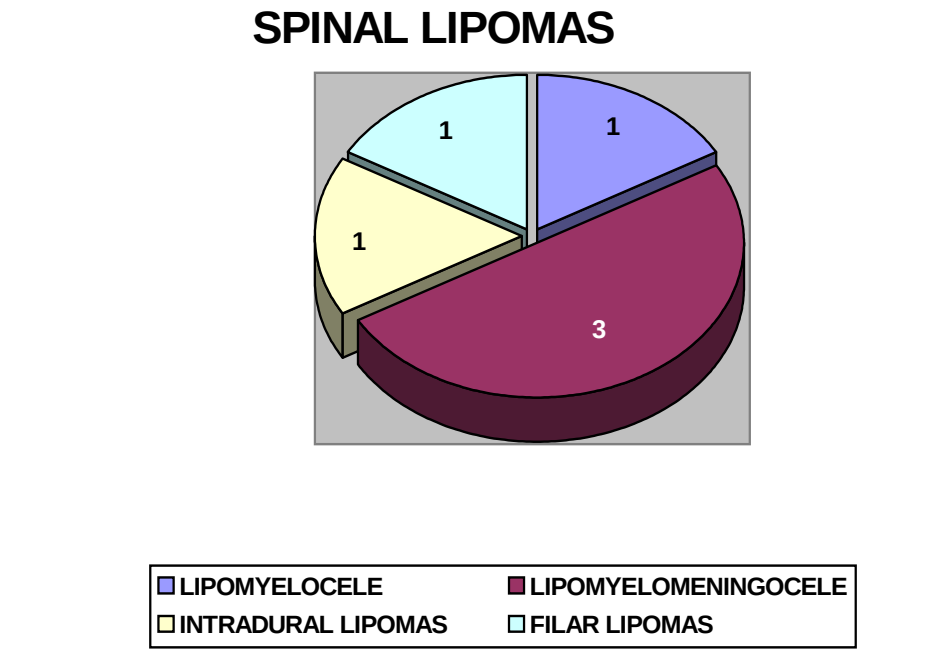

TABLE 10

DIASTEMATOMYELIA

Type	*Fibrous septum*	*Bony septum*	*Total*	*%*
Diastematomyelia in occult SD	2	2	4	28.57
Diastematomyelia in open SD	5	5	10	71.43
Total	7	7	14	100

Bony and fibrous septum occurred equally in both open and occult SD.

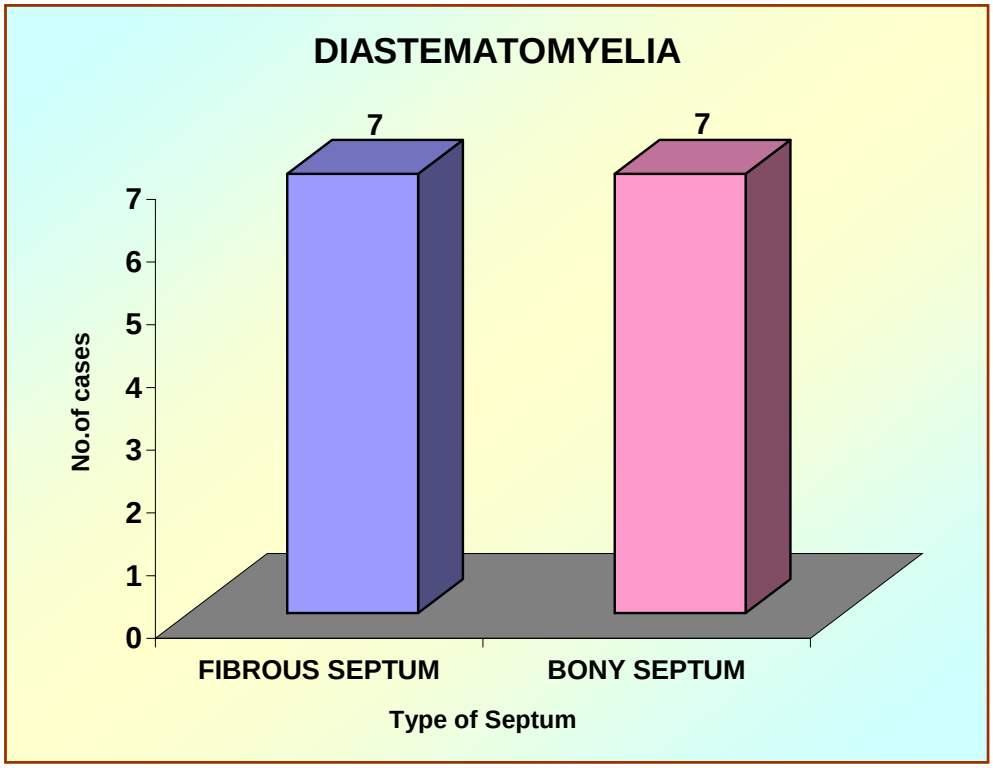

TABLE 11

DIASTEMATOMYELIA: SITES OF INVOLVEMENT IN THE SPINE

Type	*Cervical*	*Dorsal*	*Dorsolumbar*	*Lumbar*	*Lumbosacral*	*Total*	*%*
Open SD	0	1	4	3	2	10	71.43
Occult SD	0	0	2	2	0	4	28.57
Total	0	1	6	5	2	14	100

In open SD the most common site of occurrence of Diastematomyelia are DL and L regions. In occult SD Diastematomyelia occurred equally in both DL and L regions.

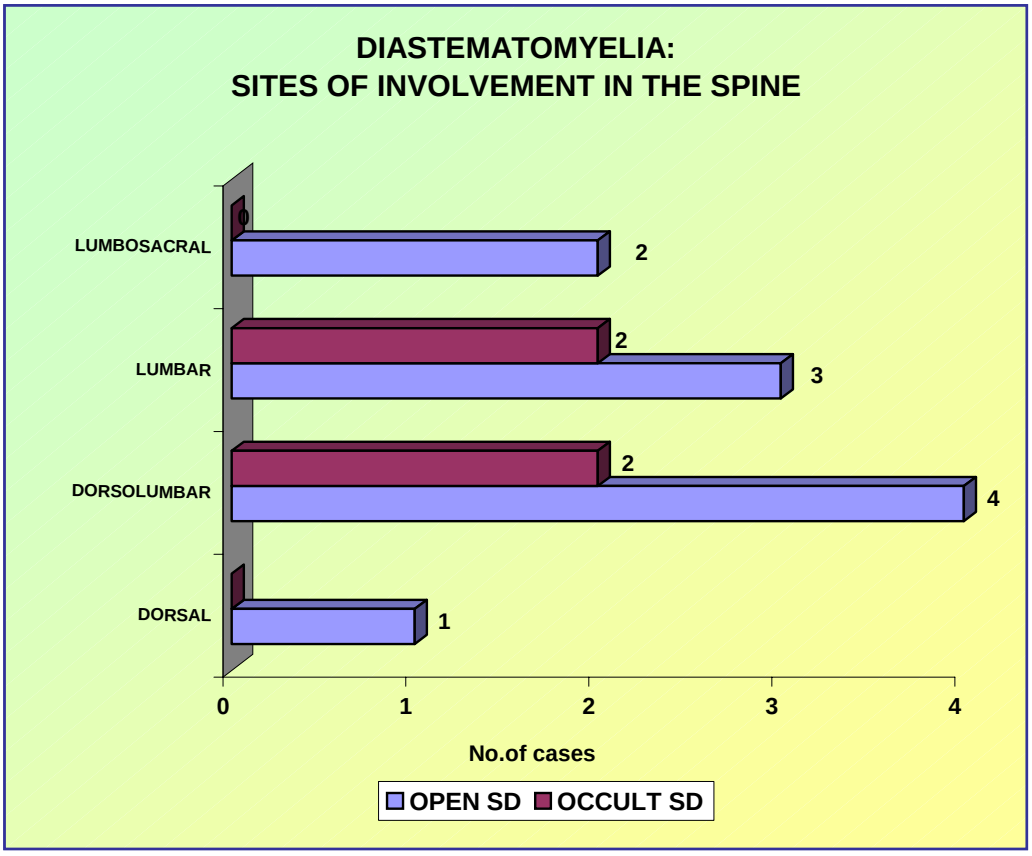

TABLE 12

TETHERING OF CORD

Type	*Tethering*	*No tethering*	*Total*
Spinal lipomas	4	2	6
Diastematomyelia	1	3	4
Open SD	4	52	56
Dorsal dermal sinus	0	1	1
Tight filum terminale	1	0	1
Anterior sacral meningocele	0	1	1
Sacral agenesis	0	1	1
TOTAL	10	60	70
%	14.29	85.71	100

Tethering of cord occurred in 14.29% of the total cases.

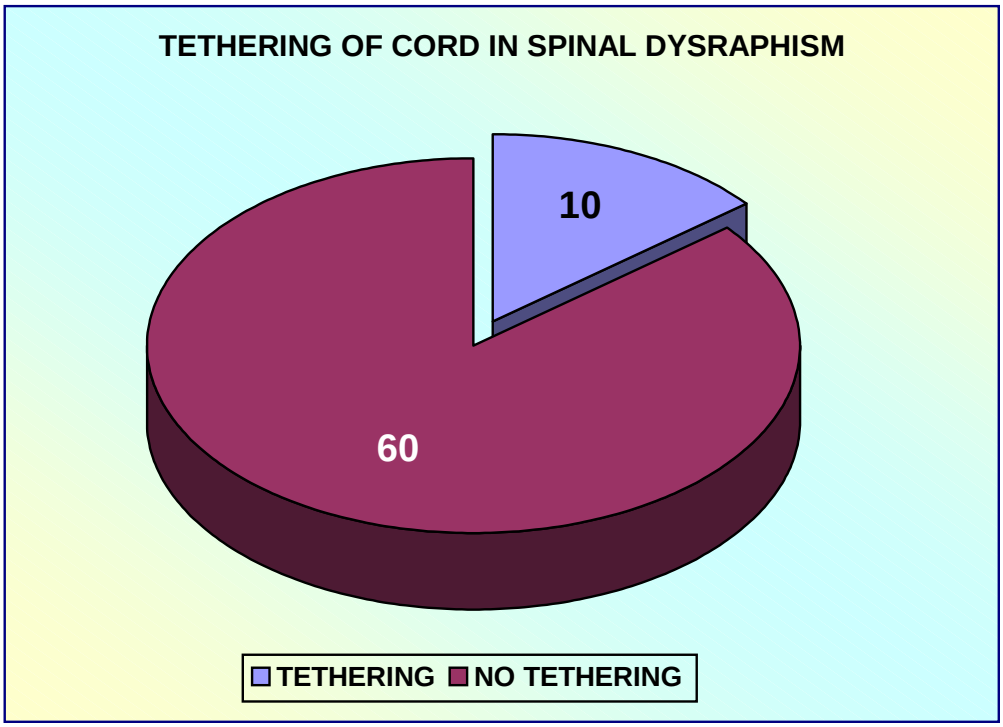

TABLE 13

VERTEBRAL ANOMALIES

	Hemivertebra	***Butterfly vertebra***	***Block vertebra***	***Spina bifida***	***Others***
Open SD	21	23	10	56	2
Spinal Lipomas	2	3	1	6	0
Dorsal dermal sinus	0	1	0	1	0
Diastematomyelia	2	0	1	3	0
Tight Filum Terminale	0	1	0	0	0
Anterior Sacral Meningocele	1	0	0	1	0
Sacral Agenesis	0	1	0	1	0
TOTAL	26	29	12	68	2
%	37.14	41.43	17.14	97.14	2.86

Spinar biffila is the most common vertebral anomaly occurring in 97.14% of total cases.

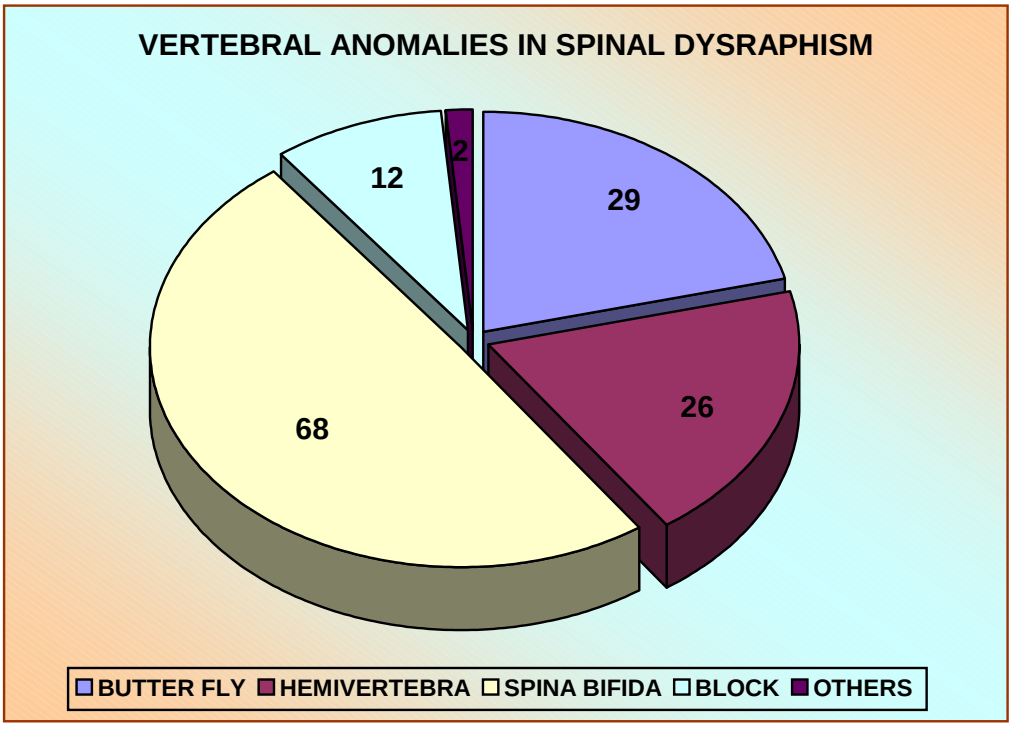

TABLE 14

SPINA BIFIDA DISTRIBUTION IN SPINE

Types	Spina bifida Cases	Distribution in spine								Total	
		Cervical		Dorsal		Lumbar		Lumbosacral			
		No	%	No	%	No	%	No	%	No	%
Open SD	56	4	7.14	12	21.43	18	32.14	22	39.29	56	100
Occult SD	12	1	8.33	2	16.67	4	33.33	5	41.67	12	100
Total	68	5	7.35	14	20.59	22	32.35	27	39.71	68	100

Spina bifida occurs in 97.14% of the total cases. Lumbosacral spine is most common site of occurrence in both open and occult spinal dysraphism.

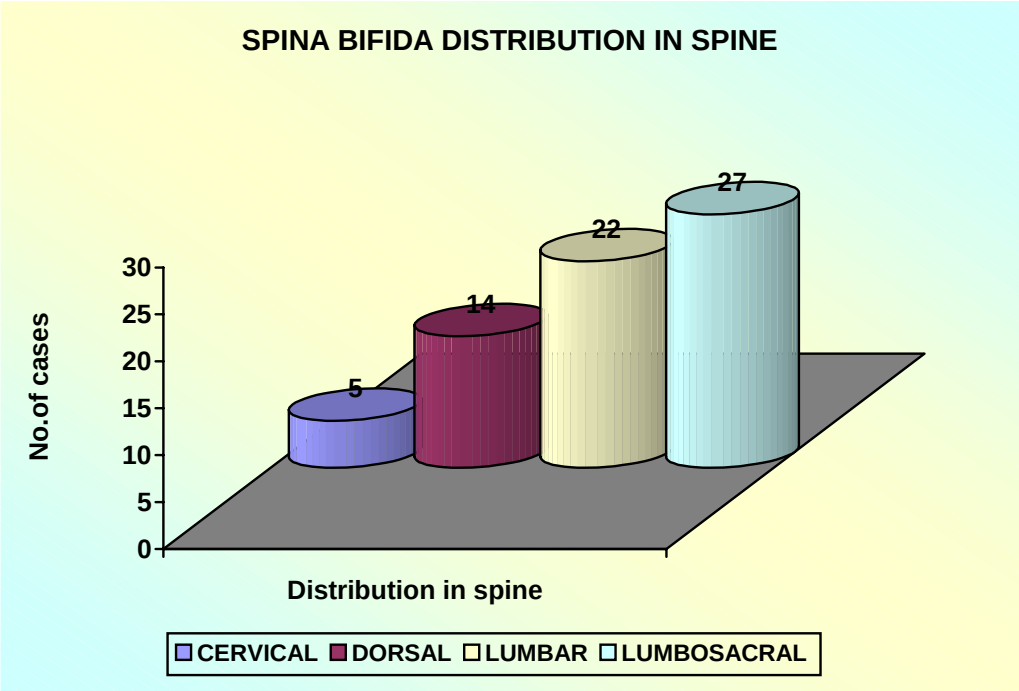

TABLE 15

DISTRIBUTION OF SPINAL DYSRAPHISM IN SPINE

Types	*No*	*Distribution in spine*								*Total*	
		Cervical		*Dorsal*		*Lumbar*		*Lumbosacral*		**No.**	**%**
Open SD	56	4	7.14%	12	21.43%	18	32.14%	22	39.29%	56	100
Occult SD	14	1	7.14%	2	14.29%	4	28.57%	7	50%	14	100
Total	70	5	7.14%	14	20%	22	31.43%	29	41.43%	70	100

The most common site of occurrence of spinal dysraphism is LS spine.

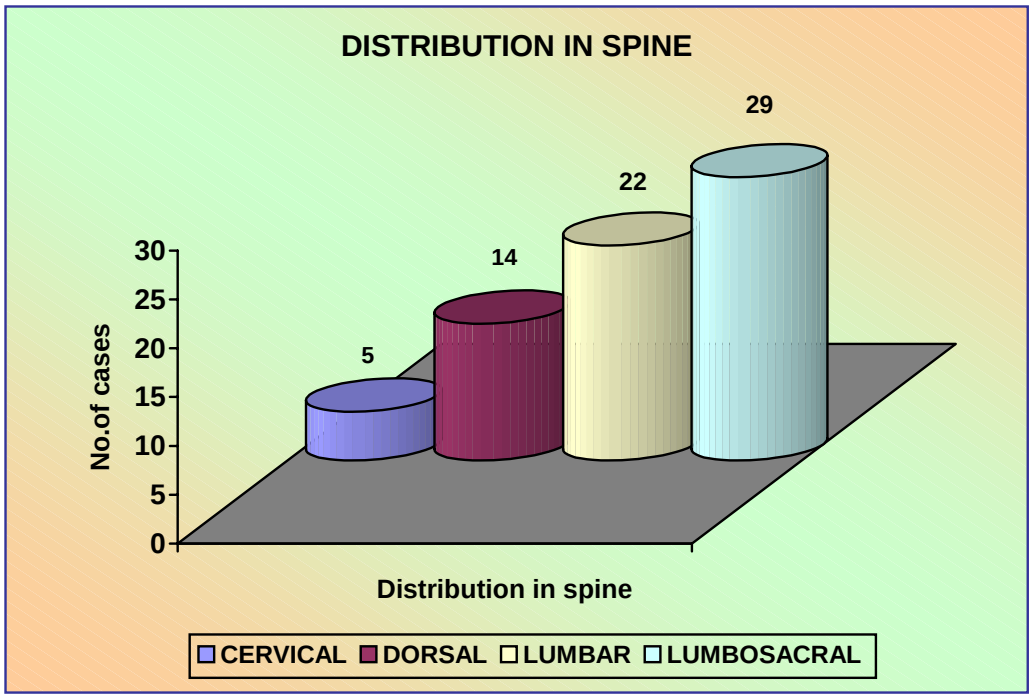

TABLE 16

SPINAL CURVATURE

Spinal curvature	*Scoliosis*			*Kyphosis*		*Lordosis*	*Total*
Region	*Cervical*	*Dorsal*	*Lumbar*	*Dorsal*	*Lumbar*	*Lumbar*	
OPEN SD	1	6	5	4	2	4	22
OCCULTSD	1	5	4	5	3	3	21
TOTAL	2	11	9	9	5	7	43
%	2.86	15.71	12.86	12.86	7.14	10	61.43

The most common spinal curvature anomaly is scoliosis.

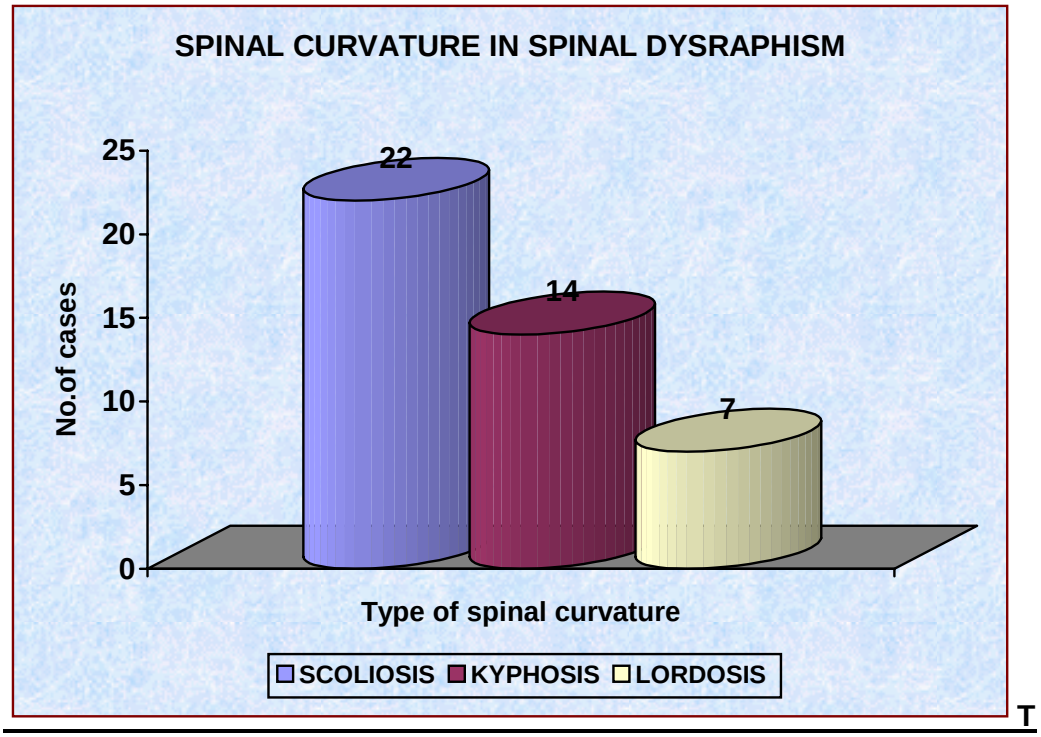

TABLE 17

HYDROMYELIA ASSOCIATION

Type	***Hydromyelia***		***Total***
	Present	***Absent***	
Open SD	15	41	56
Occult SD	7	7	14
Total	22	48	70
%	31.43	68.57	100

Hydromyelia occurred in 31.43%. It was found to be more common in open SD.

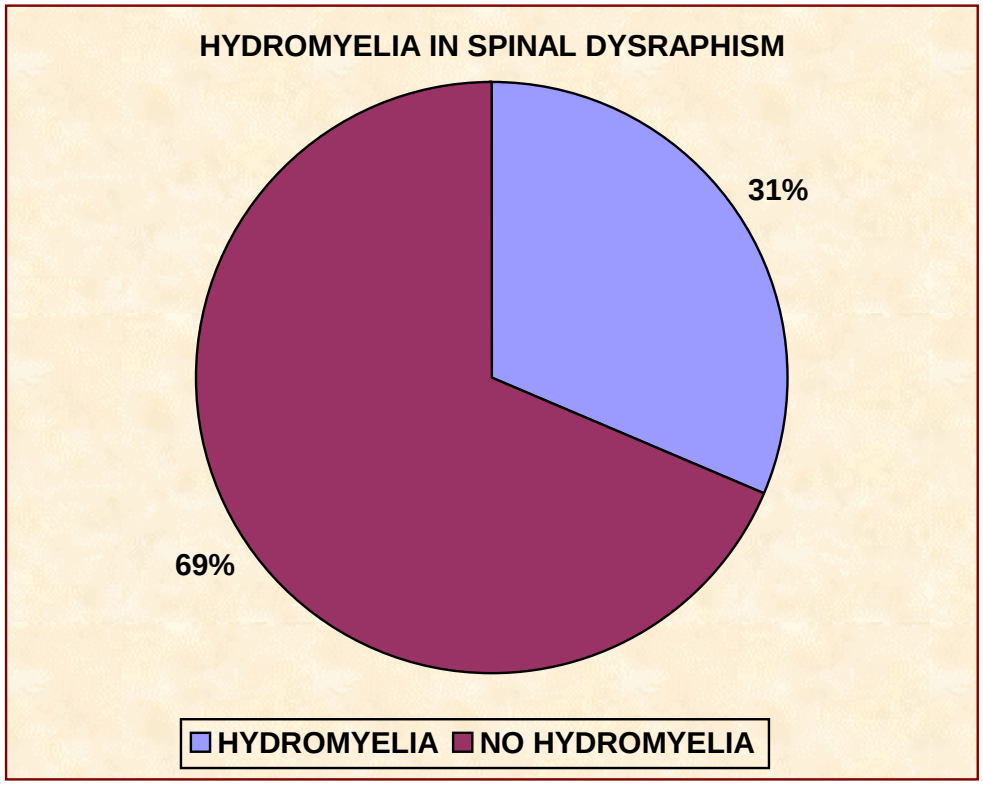

TABLE 18

HYDROCEPHALUS IN SPINAL DYSRAPHISM

Type	*Hydrocephalus*		
	Present	***Absent***	***Total***
OPEN SD	25	31	56
OCCULT SD	5	9	14
TOTAL	30	40	70
%	42.86	57.14	100

Hydrocephalus is present in 42.86% of the total cases.

Hydrocephalus is more common in Open spinal dysraphism.

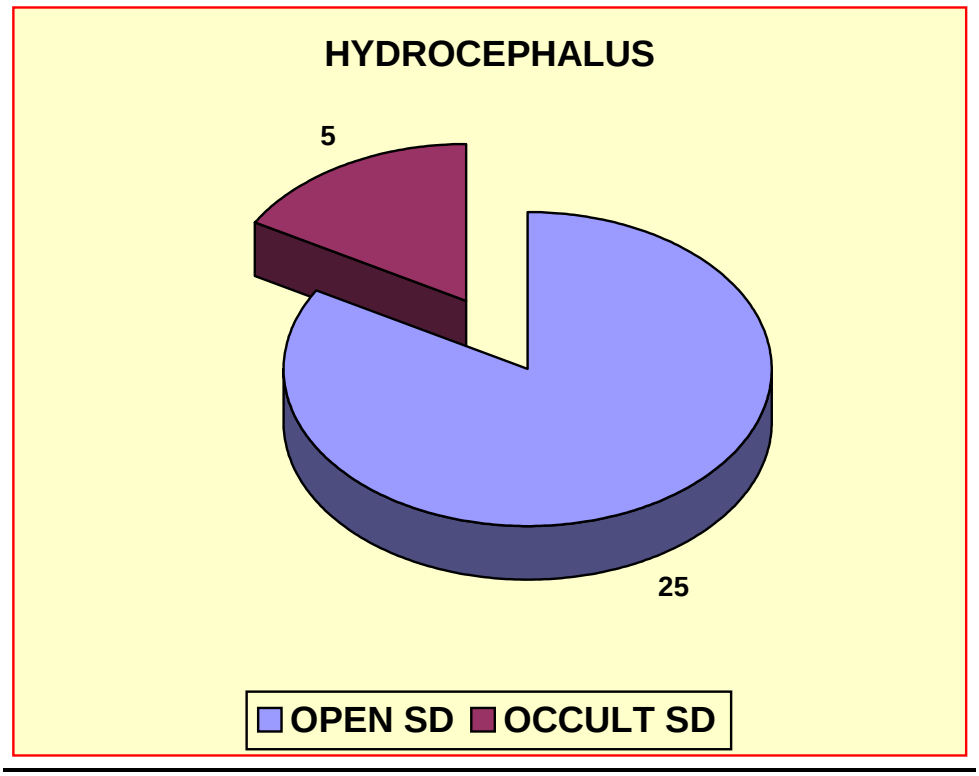

TABLE 19

CHIARI ASSOCIATION

Type	*Chiari II*	*Chiari I*	*%*
OPEN SD	51	0	91.07
OCCULT SD	0	2	14.29

Chiari II malformation is associated with open spinal dysraphism in 91.07% of cases.

Chiari I malformation is associated with occult spinal dysraphism in 14.29% of cases.

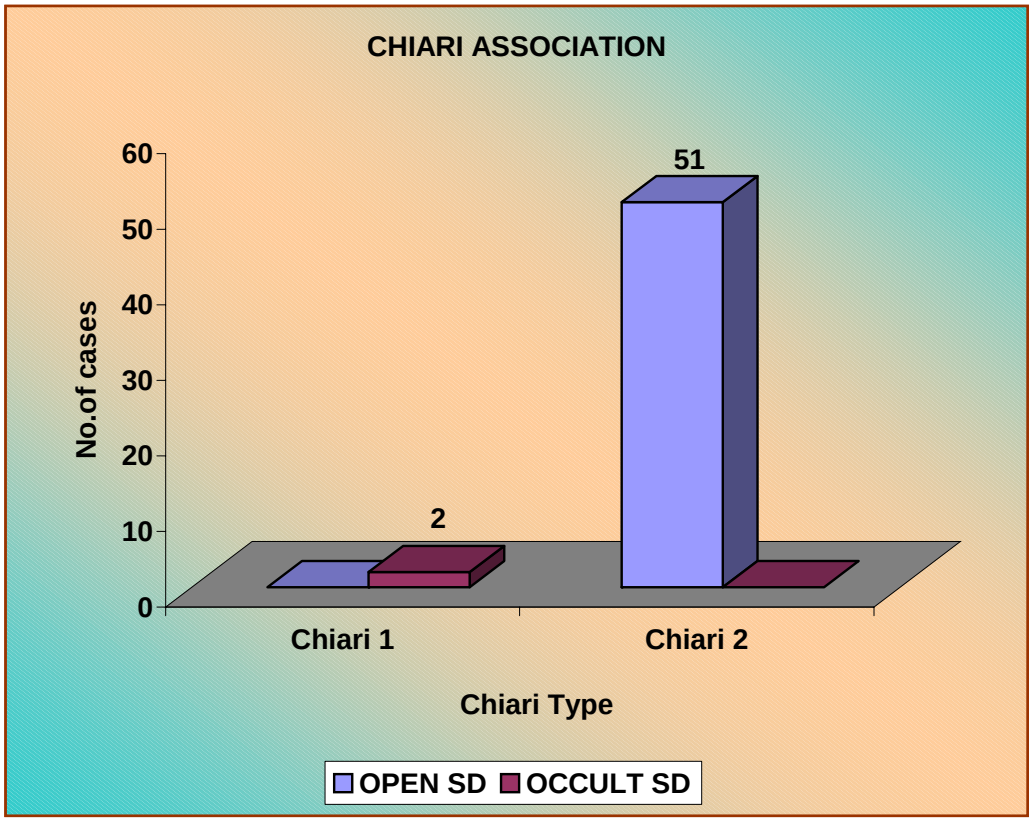

TABLE 20

COMPARISON OF CT AND MRI IN SPINAL DYSRAPHISM

S. No	*Characteristics*	*CT*	*MRI*
1	OPEN SPINAL DYSRAPHISM		
	Meningomyelocele	++	++++
	Myelocele	++	++++
	Meningocele	++	++++
2	OCCULT SPINAL DYSRAPHISM		
	Spinal lipomas	++	++++
	Diastematomyelia	++	++++
	Dorsal dermal sinus	++	++++
	Tight filum terminale	+	++++
	Anterior sacral meningocele	++	++++
	Sacral agenesis	++	++++
3	VERTEBRAL ANOMALIES	++++	++
4	DISTRIBUTION IN SPINE	+++	+++
5	SPINAL CURVATURE	+++	+++
6	TETHERING	+	++++
7	CHIARI ASSOCIATION	++	++++
8	HYDROMYELIA	++	++++
9	HYDROCEPHALUS	++	++++

KEYS

\+ POOR

++ GOOD

+++ EQUAL

++++ EXCELLENT

DISCUSSION

A total of 70 cases of spinal dysraphism were analyzed using Helical CT and MRI.

INCIDENCE

56 Patients were of open spinal dysraphism type and 14 patients were of occult spinal dysraphism accounting for 80% and 20% respectively. This is comparable with study conducted by Kumar R singh et al showing an incidence of 76.77% and 23.23% for open and occult spinal dysraphism respectively[69].

GENDER

In open spinal dysraphism there were 23 males and 33 females accounting for 41.01% and 58.93% respectively thus showing female predominance (M:F 1:1.43) comparable with the study by Steinbok P et al[54]

In occult spinal dysraphism males constituted 10 cases and females 4 cases accounting for 71.43 % and 28.53% respectively showing marked male predominance. (M: F 2.5:1). This comparable with Tripathy P et al showing male female ratio of 2.2:1[22].

AGE OF PRESENTATION

All cases of open Spinal dysraphism occurred in the first two years of life with no cases beyond that age group (Mean age of presentation is 1.23yrs). Occult Spinal dysraphism patients presented at later age group in the first, second and third decade with most of the cases occurring in the first decade. (Mean age of presentation is 6.6 yrs).

NEUROLOGICAL COMPLICATIONS

Severe neurological complications were reported in all the cases of open Spinal dysraphism. In occult Spinal dysraphism neurological manifestations were less severe and were present in 11 of the 14 cases. These findings correlate with McLone DG et al[55].

CUTANEOUS SIGNS

Among the cutaneous manifestations of Occult Spinal dysraphism most common finding was mass in the back (50%) predominantly in the Lumbosacral region followed by Dermal dimple, Hypertrichosis, Silky hair, Dermal sinus, Capillary hemangioma etc This correlates with Kumar. R. Singh et al[69]whose study showed lumbosacral swelling in 57% of cases.

OPEN SPINAL DYSRAPHISM

Among the open Spinal dysraphism the most common lesion was Myelomeningocele accounting for 53 cases out of 56 cases (94.64%) followed by Myelocele 2cases (3.57%) and Meningocele 1 case (1.79%). The study conducted by Kumar R. et al[69] showed 72%, 2% and 1% for Myelomeningocele, Myelocele and Meningocele respectively.The lesions were distributed in the Cervical, Dorsal, Lumbar and Lumbosacral regions. The Lumbosacral region was the most common site accounting for 39.29% followed by lumbar (32.14%) and dorsal (21.43%) correlating with Singh .N. et al[69]study which showed 38% for the Lumbosacral region.

OCCULT SPINAL DYSRAPHISM

Among the occult SD, Spinal lipomas accounted for 6 out of 14 cases(42.86%). The most common spinal lipoma was Lipomyelomeningocele accounting for 50% of spinal lipomas followed by Lipomyelocele, Dural lipoma and Filar lipomas all accounting for 16.67% respectively, correlating with Naidich TP et al [57]. Dorsal

dermal sinus occurred in only one case accounting for 7.14% proving that it is an uncommon lesion in our series. Kumar R Singh et al study showed 4.5% for dorsal dermal sinus.

Tight filum terminale syndrome accounted for only 7.14% in our series proving that it is an uncommon lesion among the occult SD which concurred with Fitz CR et al[59] and Love JG [60]

Among the rare caudal spinal anomalies Anterior sacral meningocele and sacral agenesis accounted for 7.14% respectively proving them to uncommon lesions in concurrence with Pang D et al[14].

DIASTEMATOMYELIA

A total of 10 cases occurred in open Spinal dysraphism (71.43%) and 4 cases in Occult Spinal dysraphism(28.57%). Fibrous and bony septum occurred equally in both types. In Open spinal dysraphism Diastematomyelia occurred most commonly in the dorsolumbar(4 cases) region followed by Lumbar(3 cases) and Lumbosacral regions(2 cases). In Occult SD Diastemetamyelia occurred equally in dorsolumbar and lumbar regions (2 cases each). These Findings Concur with Han JS et al [43]

TETHERING OF CORD

Tethering occurred in 4 cases of open Spinal dysraphism and 6 cases of closed Spinal dysraphism representing 14.29 % of the total cases. One case in the occult Spinal dysraphism represented Tight filum terminale. Tethering is associated with spinal lipomas(4 cases) and Diastematomyelia(1case) . This study correlates with N. Ragavan et al[6].

VERTEBRAL ANOMALIES

Among the vertebral anomalies Spinabifida occurred in 68 of the 70 cases representing 97.14 % as the most common vertebral anomaly followed by Butterfly

vertebra (41.43%), Hemi vertebra (37.14), Block vertebra (17.14%) and others (2.86%). concurring with study by Tripathy P et al [22]

SPINA BIFIDA DISTRIBUTION

Spina bifida was most common in Lumbosacral spine (39.71%) followed by Lumbar spine (32.35%), Dorsal spine (20.59%) and Cervical (7.35 %) concurring with the study by Roy RN et al[22], which also showed predominant involvement of spina bifida in the Lumbosacral spine.

SPINAL CURVATURE ANOMALIES

The most common spinal curvature anomaly was scoliosis (31.43%) followed by Kyphosis (20%) and Lordosis (10%). In Open spinal dysraphism, Scoliosis was most common in dorsal spine (6 cases) followed by Lumbosacral region (5 cases). Occult spinal dysrapism also showed similar distribution. In both open and occult Spinal dysraphism Kyphosis was most common in dorsal spine followed by Lumbar spine. Lordosis occurred in lumbar spine. These findings concurred with the study by Barson AJ etal[62]which also showed a predominance of scoliosis.

HYDROMYELIA

Hydromyelia was present in 22 of the total 70 cases accounting for 31.43%. 15 cases belonged to Open Spinal dysraphism while occult Spinal dysraphism comprised 7 cases. The study of Kumar R et al [69] showed an incidence of 27%

CHIARI MALFORMATIONS

Chiari II malformation occurred in 51 of the 56 cases in open Spinal dysraphism accounting for 91.07%. These findings correlate with El Gammel et al[67].

Chiari I was present in 2 cases of the 14 cases of occult Spinal dysraphism accounting for 14.29%, correlating with Naidich TP et al [68]

HYDROCEPHALUS

Hydrocephalus was present in 30 cases accounting for 42.86%, correlating with the Comparative study of complex spina bifida and split cord malformation done by Kumar Raj, Singh SN et al which showed 46%.

SUMMARY

DEMOGRAPHIC PATTERN

Open spinal dysraphism is more common than occult spinal dysraphism with a slight female predominance and presents usually in the first year of life.Almost all patients present with severe neurological manifestations.Myelomeningocele is the most common type.

Occult spinal dysraphism is less common and shows male predominance and presents at a later age group with cutaneous stigmata and minor neurological symptoms.Spinal lipomas are the most common type.Dorsal dermal sinus, Diastematomyelia and Tight filum terminale syndrome are relatively less common.Caudal spinal anomalies are rare.

ROLE OF MRI

MRI is the imaging modality of choice for characterizing the anomalies of spinal cord and associated soft tissues in spinal dysraphism.

- MRI clearly identifies and characterizes the nature of neural tissue protruding through the dysrasphic spine in Meningomyelocele,Myelocele And Meningocele.
- Spinal cord lipomas are best characterized by fat suppression sequences.

- MRI characterizes the location,extent,direction of Dorsal dermal sinus and associated Dermoid/Epidermoid.

- MRI plays an important role in the characterization of Diastematomyelia into split cord malformation I and II based on the type of septum and nature of dural covering. Size, extent and site of rejoining of the hemicords are best depicted in coronal plane. Fibrous septum is best depicted in MRI while bony septum is best recognized in CT.

- MRI is the investigation of choice in diagnosing Tight filum terminale syndrome and other causes of tethering.

- Chiari malformations, Hydomyelia and Hydrocephalus are best detected by MRI.

- Spinal curvature anomalies are well studied by MRI due to its inherent multiplanar capability.

- MRI best characterizes caudal spinal anomalies.

ROLE OF CT

CT is the investigation of choice in diagnosis of the bony anomalies of spinal dysraphism.

- Vertebral segmentation and spinal curvature anomalies are best studied by Multiplanar reformatted CT.

- Bony septum in Diastematomyelia is best diagnosed by CT.

- Meningomyelocele,Myelocele And Meningocele are diagnosed by CT but are best charecterised by MRI.

- Spinal lipomas are detected by their fat attenuation but further characterization requires MRI.

Thus CT and MRI play a complementary role in the diagnosis and characterization of various forms of spinal dysraphism and associated anomalies.

CONCLUSION

- MRI is the imaging modality of choice for evaluation of the soft tissue anomalies of Spinal dysraphism especially spinal cord anomalies.
- Multiplanar reformatted CT is an excellent imaging modality for characterization of vertebral segmentation defects, spinal curvature anomalies associated with spinal dysraphism.

Thus CT and MRI together play an important role in the complete radiological evaluation of spinal dysraphism.

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ANNEXURE

ABBREVIATION

CT	**-**	Computed tomography
MRI	**-**	Magnetic resonance imaging
T1W	**-**	T1weighted
T2W	**-**	T2weighted
FOV	**-**	Field of view
HASTE	**-**	Half fourier acquisition of single shot turbo spin echo
GRE	**-**	Gradient recalled echos
FLAIR	**-**	Fluid attenuation inversion recovery sequence
STIR	**-**	Short tau inversion recovery sequence
MPR	**-**	Multiplanar reformation
LS	**-**	Lumbosacral spine
D	**-**	Dorsal spine
DL	**-**	Dorsolumbar
C	**-**	Cervical spine
ALL	**-**	Anterior longitudinal ligament
PLL	**-**	Posterior longitudinal ligament
SAS	**-**	Subarachnoid space
CSF	**-**	Cerebrospinal fluid
CVJ	**-**	Craniovertebral junction
M	**-**	Male
F	**-**	Female

PROFORMA

STUDY ON IMAGING OF SPINAL DYSRAPHISM

Name Age/sex

Ward/unit IP.NO

COMPLAINTS

- **Lumbosacral swelling**
- **Occult signs**
  - Dimple
  - Tuft of hair
  - Nevi

- **Neurological deficits**
  - Bowel
  - Bladder incontinence
  - Motor deficit
  - Sensory deficit

- **Spinal deformity**
  - Scoliosis
  - Kyphoscoliosis
  - Kyphosis
  - Lordosis

CLINICAL EXAMINATION

Local Examination

Lumbosacral swelling

- Open
- Closed

Occult signs

- Dimple
- Tuft of hair
- Nevi

Motor System

Sensory System

IMAGING

Plain radiograph

Computed tomography

Magnetic resonance imaging

DIAGNOSIS

KEY TO MASTER CHART

SEX

1. Male.
2. Female.

OPEN SPINAL DYSRAPHISM

1. Meningomyelocele.
2. Myelocele.
3. Meningocele.
4. Diastematomyelia.
    C. Cervical.
    D. Dorsal.
    DL. Dorsolumbar.
    L. Lumbar.
    LS. Lumbosacral
5. Tethering.

OCCULT SPINAL DYSRAPHISM

1. Spinal lipoma.
2. Diastematomyelia.
    C. Cervical.
    D. Dorsal.
    DL. Dorsolumbar.
    L. Lumbar.
    LS. Lumbosacral.
3. Dorsal dermal sinus.
4. Tethered cord.
5. Meningocele.
6. Tight filum terminale.

CAUDAL SPINAL ANOMALIES

1. Caudal regression syndrome.
2. Terminal myeleocystocele.
3. Anterior sacral meningocele.
4. Occult intrasacral meningocele.
5. Sacrococcygeal teratoma.

HYDROMYELIA

1. Yes.
2. No.

HYDROCEPHALUS

1. Yes.
2. No.

CHIARI MALFORMATIONS

1. CHIARI 1.
2. CHIARI 2.

VERTEBRAL ANOMALIES

1. Spina bifida.
2. Hemivertebra.
3. Butterfly vertebra.
4. Block vertebra.
5. others.

SPINAL CURVATURE

1. Scoliosis.
2. Kyphosis.
3. Lordosis.

DORSAL ENTERIC CYST/FISTULA

1. Yes.
2. No.

OCCULT CUTANEOUS SIGNS

1. Palpable mass.

2. Dermal dimple.

3. Hypertrichosis.

4. Silky hair.

5. Dermal sinus.

6. Capillary hemangioma.

7. Rudimetary tail.

8. Atretic meninocele.

MOTOR/SENSORY DEFICIT

1. Yes.
2. No.

BLADDER/BOWEL DISTURBANCES

1. Yes.
2. No.